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Pediatric Surgery in Tropics

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Contents

In Defense of Case Reports (Editorial) *65*
Vivek Gharpure

Pediatric Gastric Volvulus in Senegal: An 8-year Review (Clinical Study) *68*
Ndeye Fatou Seck, Ibrahima Bocar Wellé, Florent Tshibwid A Zeng, Papa Alassane Mbaye, Ndèye Aby Ndoye, Aloïse Sagna, Oumar Ndour, Gabriel Ngom

Noma (Cancrum Oris) (Tropical Surgery Series - 2) *75*
Srinidhi Raveenthiran, Venkatachalam Raveenthiran

Split Notochord Syndrome (Diagnostic Image) *93*
Ketaki Gharpure

On the Role of Prophylactic Gonadectomy in Turner Syndrome (Clinical Update) *95*
Jayaprakash Sahoo, Mukesh Kumar

Removal of Double-J Stent in Children without Anesthesia or Cystoscopy: A Useful Technique (How I do it) *99*
Atreyee Sarkar, Meghna Kinjalk

A study of retroperitoneal core needle biopsy in the diagnosis of suspected malignant renal tumors (Clinical Study) *102*
Priya Singh, Yogesh Kumar Sarin, Anita Nangia

Morphology of Umbilical Hernia in Senegalese Children and Its Impact on the Clinical Presentation (Clinical Study) *108*
Gabriel Ngom, Florent Tshibwid A Zeng, Ndeye Dibor Ndiaye, Doudou Gueye, Youssouph Diedhiou, Ndèye Aby Ndoye, Papa Alassane Mbaye, Aloïse Sagna, Oumar Ndour

Posterior Urethral Valve: Is complete endoscopic valve incision sufficient? (Clinical study) *116*
Vidyanand Deshpande, Yogesh Kumar Sarin

Unraveling Noma in the Modern Age (Invited Expert Commentary) *124*
David Allen Shaye

PST PEDIATRIC SURGERY IN TROPICS

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Editorial

In Defense of Case Reports

Vivek Gharpure

Chief Editor, Pediatric Surgery in Tropics

Medical publishing began with case reports; or case series. Some doctor found an unusual patient or a unusual pattern of disease that was treated in a particular manner and thought it prudent to let others know about it. Some of the therapeutic principles which are now considered sacrosanct were established by such case reports. For example, the routine removal of inflammed appendix was established by the case series published by McBurney in 1891. He described the disease and its treatment in a paper which had twenty four patients.(1) There were no controls; no blinding; no multicentric collaboration and yet there was only one death. Other series describing such patients had up to 25% mortality. By current standards, McBurney's paper would have been rejected with a comment of 'unscientific claim'; and that would have been a bad thing to happen. He later described in detail the eponymic incision in a series of four patients.(2) Subsequently several anatomical studies found that only 1% patients have appendix located at McBurney's point and less than half of the patients with acute appendicitis have maximum pain at the point. However, it was a watershed event in abdominal surgery. One anecdotal case series of 24 patients and another series with just four patients changed the paradigm.

Not all innovations were taken in this spirit. In his book, Dr Robert Morris described an operation for relief of spinal cord compression, which was considered so dangerous by the then medical community and they complained to the sheriff. The officer was standing by to arrest Dr Morris had the patient had died.(3)

These days, most of the scholarly journals, association journals do not publish case reports or case series. Everyone wants a prospective study, a trial with controls. Open access journals do publish case reports, but only after taking a substantial fee.

Illnesses, congenital malformations have no predilection to be found only in university teaching hospitals. They can present anywhere in the world; are treated successfully by independent surgeons, who then do not find a platform to let the medical community know about their new finding.

If case reports were not published at all, or published in obscure, non-indexed journals, how will the rest of the world know about such conditions? As it is, most of pediatric surgical illnesses come under 'rare disease' category. Hence large sample randomized controlled trials are often improbable.

Case reports may stimulate an investigator to conceive hypotheses about the etiology and etiopathogenesis of the condition, which can later be tested in animal models or by clinical trials.

In neurology, almost every condition was first described as a case report. Are we to assume that all possible medical conditions, malformations,

anomalies, are already known? Thomas Kuhn, in his book 'The Structure of Scientific Revolutions', stated that all discoveries commence with awareness of an anomaly.[4] By denying case reports, editors are acting like ostriches, who bury their heads in sand, and refuse to look at new sights.

It is true, case reports do not establish principles of management, as they say, "One swallow does not make a summer". However, for the suffering patients, their illness and anomaly is of utmost importance, and if the treating doctor has no information about the management of the disorder, both are going to suffer. Case reports describing a previous occurrence, may give some guidelines, which the doctor may follow, sometimes modify according to the given situation. Some help is better than nothing. Case reports also provide others with ideas about their own patients, initiate studies to identify such condition in their patients, consider research to determine how it may be happening and make them think of other ways to treat it.

Unfortunately, the bogie of *impact factors* discourages journal editors from publishing case reports. What is an impact factor other than merely a derivative of some numbers? The correlation between circulation, citation and impact factors is tenuous at best, and depends upon myriad factors which are difficult to analyze.[5] Thus, those journals which already have a high impact factor will attract more manuscripts and may continue to have a high impact factor; while all other journals may languish. [6]

When all journals were printed, it cost much money to bring out an issue. Hence, editors of 'high impact journals' were not happy to publish case reports that occupied journal space without contributing to impact factor. Utilizing this scenario, predatory journals jumped in the fray and lured potential authors. Now, with electronic publication, the costs of publishing are minimal; there are no constraints of page number. Yet open access journals charge hefty amount to publish a two page case report!

In a parallel stream, that of social science, *'Matthew effect'* was a term coined by Robert Merton to describe as to how, among other things, eminent scientists will often get more credit than a comparatively unknown researcher, even if their works are similar. It also means that credit will usually be given to researchers who are already famous.[7] The same is perhaps true of 'high impact' journals.

Case reports are an important aspect of clinical research. When it comes to pediatric surgical illnesses, most of them come under rare disease category and numbers will be small even in big referral centers. It will be difficult to have a significant number of patients unless it is a multicentric, collaborative study. We have seen few such studies such as the MOMS (Management of myelomeningocele study), TOSCIN (Time of stoma closure in newborn), TOAST (Tracheo-oesophageal atresia study), CONTRACT (conservative treatment of appendicitis in children - a randomized controlled trial) and ICLMR (International Congenital Lung Malformation Registry). Such studies are costly, difficult to organize and are beyond the scope of individual surgeons practicing in remote rural areas of resource-poor settings.

Medicine is not mathematics, where theorems are proved, once and for all. Even in mathematics, *'Gödel's Incompleteness Theorem'* turns mathematics on its head, by proving that not everything in mathematics can be proved.[8]

The journal, *Pediatric Surgery in Tropics*, has taken a conscious decision to publish case reports, without charging any fee. The editorial team hopes, this will provide a platform for individual surgeons to express their new discoveries, however humble it may be.

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Clinical Study

Pediatric Gastric Volvulus in Senegal: an 8-year review

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Keywords
Gastric volvulus
Diagnostic delay

Abbreviations
GV - Gastric Volvulus
UGICS - Upper gastro-intestinal contrast series

Abstract

Introduction: *Gastric volvulus is a rare condition in children but is infrequently reported from sub-Saharan Africa.*

Methods: *We conducted a two-center retrospective study over 7 years (2012 – 2019) from Dakar, Senegal.*

Results: *Our study included 10 patients, the age ranging from 1 month to 11 years, without any sex predominance. Six patients had a chronic presentation. Non-bilious vomiting was found in all patients. Abdominal plain X-ray (n=10) and upper gastrointestinal series (n=7) were the most frequently requested investigations. Duration of symptoms ranged from 1 day to 2 years in patients with chronic presentation and from 1 to 4 days in those with acute manifestations. Mesentero-axial type was found in 6 cases, with an associated diaphragmatic hernia in 3 cases. An associated anomaly was found in 6 patients. In all cases, the treatment consisted of open surgery with gastropexy and adjunctive surgical treatment in patients with an associated anomaly. The postoperative outcome was unremarkable in 9 cases, and 1 patient presented with recurrence of left Bochdaleck hernia after 12 months of follow-up.*

Conclusion: *Diagnosis and management of gastric volvulus was delayed in sub-Saharan Africa. Open surgery gives favorable outcomes even in resource-limited setting.*

INTRODUCTION

Gastric volvulus (GV) is known to occur in the setting of lax ligamental support of the stomach with resultant abnormal rotation. It is a rare surgical emergency, with 10 to 20% occurring in children, of whom 58% present before the first year of life.[1,2] In this the stomach may get twisted more than 180° either on long-axis (organo-axial) or short-axis (mesentero-axial) or both.[1] The presentation may be acute or chronic, depending on the duration of symptoms. The cardinal features of GV include the triad of abdominal pain, intractable retching, and inability to pass a

nasogastric tube into the stomach. Sometimes, non-specific symptoms may lead to a delay in the diagnosis, thus worsening its prognosis.[3,4]

The main complication of GV is gastric perforation due to gastric wall necrosis. Given the life-threatening nature of GV, it should be suspected even in cases of atypical occlusive symptoms.[4] African studies on this topic are scarce.[5] Our study aims to report the diagnostic and therapeutic aspects of GV highlighting the practice in sub-Saharan Africa.

PATIENTS AND METHODS

We conducted an eight-year retrospective cross-sectional study at Albert Royer National Children's Hospital Center and Aristide Le Dantec Hospital in Dakar, Senegal. Although there are 2 other pediatric surgical services (in Diamniadio and Kaolack), these 2 services are the only public pediatric surgical facilities in Senegal.

We reviewed data from January 2012 to December 2019, which included all patients diagnosed with GV alone (primary GV) or GV associated with a coexistent pathology (secondary GV). Patients whose medical files had insufficient data were excluded. Information was collected from both medical files and operating room registers.

Collected variables were age, sex, referral diagnosis, time from symptoms to presentation at our services, clinical findings, imaging results, clinical presentation, time to surgery, type of surgical management, and outcomes. Data were saved and analyzed using Excel software.

RESULTS

Our two services identified 10 cases during the study period. The patients' ages ranged from 1 month to 11 years. There were 5 boys and 5 girls. Nine of the 10 patients previously attended another medical facility before reaching our hospital: 6 attended one medical facility, 2 attended two facilities, and 1 attended three facilities. Of the 9 referred patients, 6 had no referral diagnosis, 2 were correctly diagnosed with GV and 1 was incorrectly diagnosed as hypertrophic pyloric stenosis. The latter was 11 months old.

Duration of symptoms ranged from 1 day to 2 yrs in chronic GV and 1-4 days in acute GV. Six patients had chronic GV and 4 had acute GV. Symptoms included non-bilious vomiting in all patients, abdominal pain (n=6), and bloating (n=4). Physical examination revealed abdominal distension in 4 cases that was predominantly epigastric in 3 cases. Three patients presented with respiratory signs; of whom severe respiratory distress was found in a single case. In 2 patients, weight loss was reported. The most commonly requested imaging investigation was the plain thoraco-abdominal X-ray (n=10), followed by upper gastrointestinal contrast series (UGICS) (n=7) (Fig. 1), computed tomography (n=3), and abdominal ultrasound-Doppler scan (n=1).

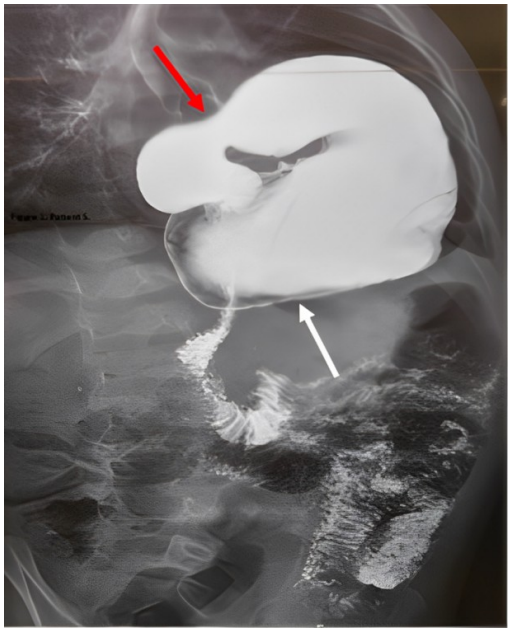

Fig 1. Mesentero-axial gastric volvulus

Upper gastrointestinal contrast radiograph of the patient-5 depicting the pylorus in the left upper abdomen (red arrow) with an inverted fundus (white arrow).

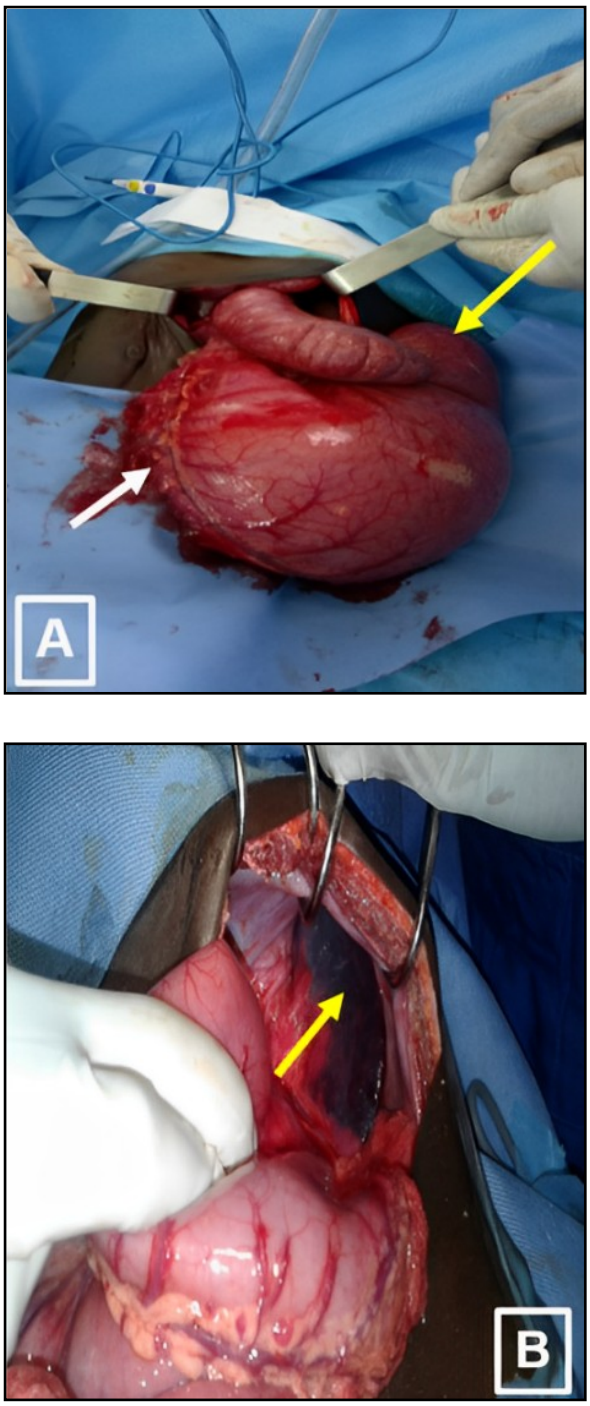

Fig 2. Intraoperative findings in patient-1.
(A) - Mesentero-axial gastric volvulus with the pylorus in the left upper abdomen (yellow arrow) and the fundus in the lower position (white arrow). (B) - Left Bochdaleck hernia involving the spleen (yellow arrow).

The diagnosis of gastric volvulus was made pre-operatively in 7 cases. The type of gastric volvulus was specified in 8 cases: mesentero-axial in 6 and organo-axial in 2. In 2 patients, spontaneous derotation was noted on surgical exploration; hence the type of volvulus could not be ascertained, as UGICS series had not been performed in these cases. In all cases, the standard anatomical anchors of the stomach were absent. In 6 patients, associated anomalies were present. (Table 1)

The time from the first consultation in one of the two pediatric surgical departments to the surgery date ranged from 4 days to 4.5 months in patients with chronic gastric volvulus and from 1 to 11 days in patients with acute gastric volvulus. All patients underwent laparotomy by upper abdominal transverse incision. The treatment consisted of gastropexy in all the patients. Adjunct surgical treatment included diaphragmatic hernia repair in 2 cases (Fig. 2), diaphragmatic plication, hiatus hernia repair, Nissen fundoplication, appendectomy, removal of supernumerary spleen, and gastric perforation suture (Fig. 3), each in one case.

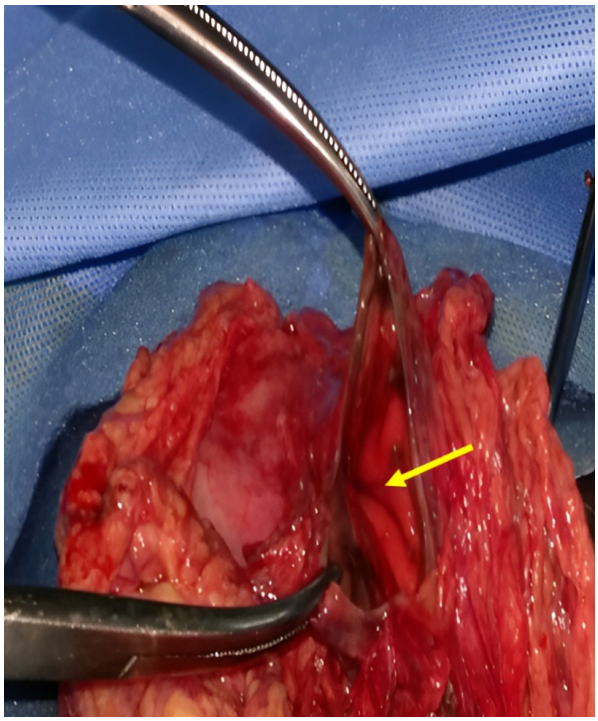

Fig 3. Intraoperative findings in patient 6
He had a history of trauma. Posterior gastric perforation is shown (yellow arrow)

Table I. Summary of clinical details, diagnostic aspects and outcome of gastric volvulus

Pt.No.	Sex	Age	Symptoms (duration)	Imaging	Volvulus type	Associated anomalies	Follow-up	Outcome
1	M	4 yr	AP, Vomiting, constipation (4d)	XR, Doppler	Acute MAV	Left CDH	24 m	Unremarkable
2	F	11m	AP, Vomiting (3m)	XR, UGICS	Chronic MAV	Left CDH	15 m	Unremarkable
3	F	12m	Vomiting (6m)	XR, UGICS	Chronic GVUS	Nil	4 yr	Unremarkable
4	F	17m	AP, AD Vomiting (8m)	XR, UGICS, CT	Chronic MAV	Left CDE, Wandering spleen	4 yr	Unremarkable
5	F	4 yr, 3 m	AP, AD, Vomiting (1d)	XR, UGICS	Acute MAV	Accessory spleen	3 m	Unremarkable
6	M	8 yr	AP, Vomiting, Hematemesis (2d)	XR, CT	Acute MAV	Left CDH, Gastric perforation, Splenic and colonic contusions	19 m	Recurrence of Bochdaleck hernia
7	M	2 yr, 7 m	Vomiting (4 m)	XR, UGICS, CT	Acute OAV	Hiatus hernia	3 m	Unremarkable
8	M	1 m	Vomiting, AD (2 wk)	XR, UGICS	Chronic MAV	Nil	3 m	Unremarkable
9	F	1 m	Vomiting (few hrs)	XR, UGICS	Chronic OAV	Nil	4 m	Unremarkable
10	M	11yr	AP, AD, Vomiting (2 yr)	XR	Chronic GVUS	Nil	3 m	Unremarkable

AD - abdominal distension; AP - abdominal pain; CDE - Congenital Diaphragmatic Eventration; CDH - Congenital Bochdaleck Diaphragmatic Hernia; CT - Computed Tomography; GVUS - Gastric volvulus (unspecified); MAV - Mesenteroaxial volvulus; OAV - Organoaxial volvulus; UGICS - Upper gastrointestinal contrast series; XR - abdominal plain x-ray

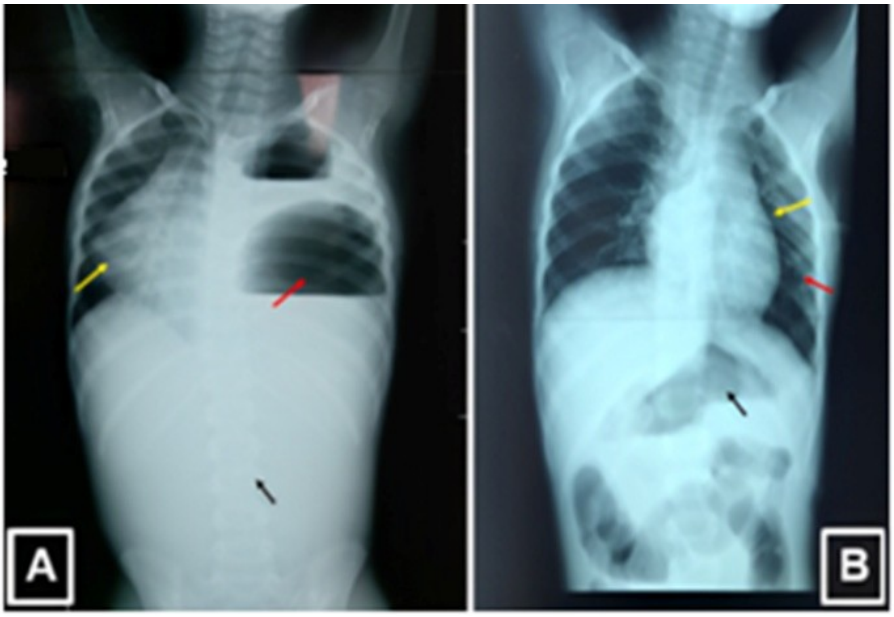

Fig 4. Gastric volvulus associated with left Bochdaleck hernia.

(A)-Preoperative radiograph showing intra-thoracic stomach (red arrow) with right-sided mediastinal shift (yellow arrow) and gasless abdomen (black arrow). (B)-Postoperative radiograph on day-30 showing full recovery of the left lung (red arrow), return of the mediastinal to normal position (yellow arrow), and presence of bowel gas (black arrow).

All patients had unremarkable early postoperative outcomes except one child. He was an 8-year-old boy who presented with an acute post-traumatic gastric volvulus associated with a left Bochdaleck hernia. He was treated with gastric detorsion and reduction of the hernia contents but without diaphragmatic repair because the defect was more than 50% of the hemi-diaphragm. The treatment of diaphragmatic hernia was delayed due to non-availability of synthetic patches necessary to close of the significant defect. On delayed extubation at 24 hrs, epigastric distension persisted and abdominal pain was marked. This prompted surgical re-exploration one day after the first operation. On re-exploration perforation of the posterior gastric wall was noted. He underwent repair of the diaphragmatic hernia without a synthetic patch and suturing of the gastric perforation. The early outcome of this second operation was unremarkable.

The follow-up period ranged from 3 months to 4 years. Nine patients had unremarkable recovery without any long-term sequelae. (Fig. 4) In the patient with a post-traumatic gastric volvulus associated with a left Bochdaleck hernia, a recurrence of the diaphragmatic hernia was noted after 12 months and a repair with a patch was planned

DISCUSSION

With a frequency of 1.4 cases per year, GV is a rare condition in our daily practice. The Senegalese literature has reported two cases of GV in adults.[6,7] A large pediatric study reported 150 cases of GV from Senegal.[8,9] The diagnosis is rare in our setting, and this can be attributable to the lack of awareness about GV among the referring doctors in peripheral health facilities. Thus, only 2 children were correctly diagnosed before their admission to our services. The rarity of this pathology is also partly due to the robustness of e gastric ligaments, which reduces its mobility.[10]

In our study, as well as in the literature, non-bilious vomiting, abdominal distension and abdominal pain were the common symptoms of GV. The classic Borchardt's triad (unproductive retching, epigastric pain or distention and the inability to pass a nasogastric tube) is rarely encountered in children.[3]

The diagnosis of GV is often challenging since the symptomatology varies in acute or chronic presentations.[2] In our study, the delay between the first consultation in one of the pediatric surgical services and the surgery date was eight times shorter for acute presentations. This difference is explained by acute gastric volvulus leading to a sudden onset of upper gastro intestinal obstruction with epigastric distension. Symptomatology of chronic gastric volvulus is insidious. It evolves through periodic exacer-bations and remissions along with non-specific symptoms such as non-bilious vomiting, respi-ratory signs, and growth retardation.[11,12] This atypical symptomatology, combined with limited knowledge of the disease and low index of suspicion in our settings, makes GV a difficult and challenging diagnosis.

Abdominal plain X-ray seems sufficient for diag-nosing acute forms by showing the classical double-bubble sign.[13] Other authors suggest that the CT scan should be the first-line examination.[9] Considering the limitations of our setting, upper gastrointestinal contrast study appears to be the critical tool for confirming GV and its type, as CT is sometimes unavailable in an emergency. Some authors reported that the UGICS can identify up to 82% of gastric volvulus and hence is the preferred investigation.[2,13-15] In our experience, it helped to diagnose GV in 7 out of 10 patients.

In our study, diaphragmatic hernia is the most frequently associated condition, in which abnormal attachment of gastric ligaments leads to intra-thoracic gastric ascension. The acute type of GV is often associated with congenital anatomical abnormalities, the most common of which are diaphragmatic hernia, diaphragmatic eventration, para-esophageal (hiatus) hernia, and congenital

asplenia or wandering spleen.[2,16] Our series noted an exceptional hiatus hernia associated with a chronic volvulus. Two patients with an acute GV had an associated diaphragmatic hernia, and one had an accessory spleen. Chronic and acute types also differ in their etiology. Generally, the chronic form is idiopathic.[11] In our series, 4 of the 6 patients with chronic GV had no associated anomaly. Authors rarely reported associated anomalies in chronic volvulus.[11] Indeed, chronic GV is caused by simple laxity of the gastric ligaments. The absence of an organic cause could explain spontaneous resolution of episodic volvulus in these patients and therefore the insidious mode of evolution by intermittent flare-ups.[11]

The surgical management requires a gastropexy and repair of the diaphragmatic defect, if a diaphragmatic hernia is associated.[11] All patients underwent a laparotomy repair because minimal invasive surgery is still not well established in our country. Open surgery allows the reduction of intra-thoracic GV with performance of additional procedures like repair of diaphragm.[17] Therefore, it appears to be more suitable for acute GV.

Despite the increasingly satisfactory results of conservative treatment in chronic forms,[11] all our patients have benefited from surgical management. This approach appears safer in African context, where the required follow-up of conservative treatment may not be possible due to lack of parental understanding of the disease and financial constraints.

The mortality rate in acute GV is twice that of chronic GV.[2] This is due to the possibility of ischemia and perforation occurring in 5-28% of cases.[2] No death was recorded in our study, but only one case of gastric perforation was found in a patient with an acute GV. Therefore, it is a presentation with high morbidity and mortality that deserves the attention of practitioners.

CONCLUSION

The diagnosis and management of GV was delayed in our set-up, especially in its chronic form, due to lack of awareness among referring doctors. The UGICS was the core radiological diagnostic examination, which should be considered in case of atypical gastric outlet obstruction. Open gastropexy resulted in a favorable outcome even in resource-limited setting.

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Tropical Surgery Series - 2

Noma (Cancrum Oris)

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Keywords
Noma
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Facial reconstruction

Abbreviations
ANG - Acute necrotizing gingivitis
CMV - Cytomegalovirus
FN - Fusobacterium necrophorum
HIV - Human immunodeficiency virus
LIMC - Low-middle-income countries
PEM - Protein energy malnutrition
TMJ - Temporo-mandibular joint
WHO - World Health Organization

Abstract

Noma (also known as cancrum oris, orofacial gangrene of childhood) is a synergistic infection of the oral cavity that causes mutilating necrosis of the orofacial soft tissues and bones. Classical noma, noma-like disease and noma neonatorum are the three clinical variants. Fusobacterium necrophorum, Prevotella intermedia, Peptostreptococcus and Pseudomonas are the key organisms incriminated in the pathogenesis. It usually starts as acute necrotizing gingivitis and progresses to necrotizing periodontitis, necrotizing stomatitis, necrotizing fasciitis and osteo-myonecrosis in that order. Noma is a disease of poverty. Synergistic bacterial infection, immuno-suppressed state and mucosal ulceration form the etiopathogenic triangle. The region extending between Senegal and Ethiopia is known as 'noma belt' due to very high incidence. W.H.O classified noma into 5 clinical stages. Acute noma is managed by giving antibiotics and restoring oral hygiene. The residual deformity of noma offers great technical challenge to the reconstructive surgeon. Facial reconstruction should be individualized based on the nature of tissue defect. With proper treatment the current mortality is less than 10%. In 2023 W.H.O announced noma as a 'neglected tropical disease' and declared that 'noma is more than a disease'.

INTRODUCTION

Noma is a synergistic infection of the oral cavity that causes mutilating necrosis of the orofacial soft tissues and bones.[1-6] It is nicknamed as 'the face of poverty'.[7] It is common in low, middle-income countries (LMIC) among malnourished children of age 1-6 years.[3,5,6,8] It is not clear as to how noma is pathologically different from other necrotizing fasciitis such as Fournier gangrene and Meleney ulcer.[9] Perhaps, destruction of underlying bone is unique of noma. Despite being known for over 1300 years, etiopathogenesis of noma is still elusive.

Acquiring scientific knowledge and reliable statistical data of noma are precluded by several socioeconomic factors.[10] As it is quickly fatal,

several patients die in remote villages even before getting medical help. Social stigma, poverty, poor transport facilities, illiteracy and distrust with allopathic medicine prevent affected children from attending hospitals. Native medicine and witchcraft further worsen the situation. As noma is common among nomadic populations proper enrollment, statistics keeping and long term follow-up are often impractical. Resource limitations in endemic areas preclude sophisticated investigations and advanced surgical corrections.

HISTORY AND ETYMOLOGY

The term *noma* was derived from the Greek word *'numein'*, meaning *'to devour'* or *'to graze'*.[11]. Ancient Greek physicians like Hippocrates (circa 460 - 377 BCE) and Galen (circa 129 – 216 CE) used this term to mean any rapidly progressing disease and they never specifically described orofacial gangrene.[11] Jan Yperman (circa 1260 – 1331 CE), a Flemish surgeon, was the first to mention *'water canker'*, the old Dutch name for noma.[12] The word *'canker'* simply means '*fungating ulcer*' and had been applied to a variety of pathologies including fungal granuloma and neoplasia. In 1595, Carolus Battus gave the first clear description identifying it as a separate entity. In 1649, Arnoldus Boot coined the term *'cancrum oris'*. However, in 1826, Coates criticised the word *'cancrum'* as *"an odd grammatical blunder"* because it uses accusative case *'cancrum'* instead of the proper nominative case *'cancer'*.[13] Unfortunately, until the 18th century, the terms '*canker oris'* and '*cancrum oris'* were confusingly used to mean both oral cancers and orofacial infective necrosis. In 1680, Cornelis van de Voorde was the first to suggest exclusive usage of the term 'noma' to mean orofacial gangrene of childhood.[11]

In 1765 Gabriel Lund recognized the 4 stages of the disease.[7] At the same time, Symmonds elucidated the etiological relationship between noma and measles. In 1781, Leendert Stelwagen was the first surgeon to do successful surgical correction of noma.[7] In 1828 Adolph Richter published the first monograph on noma.[7] In early 19th century Esmarch devised mandibular osteotomy for treating trismus and Estlander described his technique of lip-sharing reconstruction.[7]

With the advent of microbiology as a separate field, Stewart of Leeds, in 1912, isolated a spiral organism (*'Bacillus nomae'*) and concluded noma as a disease of opportunistic infection.[7] This organism was later named as *Borrelia vincentii* and subsequently lost its nominal identity due to frequent taxonomic revisions. During the World War-II, in the concentration camps of Germany and Japan, a noma-like disease (also known as *trench fever* or *Vincent disease*) was described.[14] Subsequently, a distinction between *Vincent disease* and *noma* was drawn for a brief period. Vincent disease (also known as *acute necrotizing gingivitis*) is now once again considered as the prodromal lesion of noma.[14] Interestingly, until the last century the term 'noma' was used to denote genital gangrene in young girls (*noma pudendi*).[7,12]

With the discovery of antibiotics, survival of noma greatly improved. Those who would have died of sepsis, began to live long. This necessitated the shifting of focus from sepsis control to surgical reconstruction of the survivors. Michael Tempest is considered as the founding father of modern noma surgery.[7]

Despite its existence over a millennium, noma has largely been ignored by the western researchers. Finally, World Health Organization (WHO) announced noma as a *'disease of high priority'* in 1994 and as a '*neglected tropical disease'* of great importance in December 2023.[15] WHO slogan is *"Noma is more than a disease"*.[15] GESNOMA (Geneva Study Group on Noma), a humanitarian voluntary association intended to study the pathogenesis of noma, was launched in 2001.[16] In the International Classification of Diseases (ICD-10) noma is coded under '*A69.0: Necrotizing Ulcerative Stomatitis'*.

Box 1: Synonyms of Noma ‡

ANG	Mund krebs
Acute ulcerative gingivitis	Necrotic gingivitis
AUMG	NUS
Aka popo (Zambia)	Orofacial gangrene
Anarchie dentaire	Pagnad Pak Poue (Laos)
Aphthae serpentes	Periodontal gingivitis
Bakin Kare † (Nigeria)	Periodontal stomatitis
Cancer aqauticus sive aquous §	Phagedenic gingivitis
Cancer scorbuticus labii	Plaut-Vincent stomatitis
Cancrum oris infantium	Putrid stomatitis
Canker oris	Putrid mouth sore
Can tan ma (Vietnam)	Schwartzer Krebs
Cheilocace maligna	Scorbutic canker
Ciwon Iska * (Nigeria)	Septic gingivitis
Cone gangreneux	Stomatite charbonnause
Diphtheroid angina	Stomatitis epidemiea
Feldzug stomatitis	Stomatitis gangrenosa
Fetid stomatitis	Stomatitis ulceromembranacea
FS marginal gingivitis	Stomatitis ulcerosa
FS periodontal gingivitis	Stomatocase gangraenosa maligna
Fusospirochetal gangrene	Stomatomalacia putrida
Fusospirochetal gingivitis	Trench mouth / gums
Fusospirochetosis	Todtenwurm
Gangrena oris	Ulcerative gingivitis
Ganrena accutissima caseosa pulpa	Ulcerative stomatitis
Gangraenopsis	Vincent angina
Gangrenous stomatitis	Vincent disease
Gangrenous osteo-gingivitis	Vincent gingivitis
Gilmer's disease	Vincent infection
Mouth canker	Vincent periodontitis
Mund faule	Vincent stomatitis
	Water canker §
	Wasser krebs §

‡ *Compiled from Talma[7], Marck[7], Murayama.[22] This list also contains synonyms of ANG because it is included as Stage-1 Noma in WHO classification.*

* *A Nigerian term meaning 'Disease of the wind'*

† *A neologism meaning 'Dog mouth' coined for the purpose of public awareness by radio announcements in Nigeria.*

§ *These names suggest spread of infection by contaminated water.*

ANG-Acute necrotizing gingivitis; AUMG-Acute ulcero-membraneous gingivitis; FS-Fusospirallary; NUS-Necrotizing ulcerative stomatitis

NOMENCLATURE

Noma is known by a variety of scientific and colloquial names (Box 1).[17] Three distinct clinical forms are recognized: Classical noma, Noma-like lesions and Noma neonatorum.[2] *Classical noma* is due to synergistic infection of aerobic and anaerobic bacteria occurring in malnourished children. Morphologically similar, but pathologically less aggressive lesion occurring in immuno-compromised or debilitated adults and experimental animals is called *'noma-like lesion'*.[18] In 1976 Ghosal introduced the term *'noma neonatorum'* to denote orofacial necrosis due to *Pseudomonas* occurring in edentulous premature neonates.[19,20] It was also suggested that noma neonatorum could be a form of *ecthyma gangrenosum*.[21] Although clinical manifestations are grossly identical between the 3 entities, they significantly differ in pathogenic mechanisms and outcomes. (Table 1)

ETIOPATHOGENESIS

Noma is generally considered to be a synergistic infection of aerobic and anaerobic bacteria.[24,25] Facultative aerobes thriving beneath dental plaques secrete proteolytic enzymes that cause local cell death and ulceration. Dead tissue with reduced oxidative stress facilitates growth of anaerobes. Cytotoxins (hemolysin, endotoxin and leukotoxin) secreted by anaerobes offer immunological protection to aerobes by destroying phagocytic neutrophils. Hemin, an iron containing growth factor essential for aerobes, is also released from hemoglobin by the action of hemolysin.[24] Thus, a synergistic alliance and vicious cycle is established culminating in extensive tissue damage.

Some of the anaerobes are also independently capable of creating their own favorable hypoxic micro-environment by inducing small vessel thrombosis.[26] For example, *Fusobacterium necrophorum* (FN) secretes hemagglutinin and platelet agglutinin that cause vascular thrombosis, dermonecrotic toxin that causes necrosis of the

Table 1: Clinico-pathological differences between the 3 clinical types of noma

	Classical noma	**Noma-like lesion**	**Noma neonatorum**
Nature of infection	Synergistic polymicrobial	Non-specific opportunistic infection	Monomicrobial
Common organisms	*Prevotella* and *Fusobacterium*	Opportunistic bacteria or fungi	*Pseudomonas aeruginosa* (86%)
Source of infection	Community acquired	Community or hospital acquired	Hospital acquired
Origin of infection	Interdental plaques	Hematogenous	Hematogenous
Precursor lesion	Necrotizing gingivitis	None	None
Geographic pathology	Common in LIC	Anywhere including HIC	LMIC
Affected age group	1-6 yrs	Older children, Adults	Newborn
Predisposing condition	Malnutrition, Measles	HIV, Cancer chemotherapy	Prematurity
Metastatic gangrene	Seldom	Never	Genito-perineal involvement in 20%†
Surgical challenge	Huge tissue deficit, malnutrition	Underlying immuno-compromised state	Seldom reach the stage of surgical reconstruction
Clinical course	Rapidly fatal within a few days/weeks	Indolent with survival for several weeks/months	Rapidly fatal within few days
Current Mortality of acute phase	8-15%	54 - 78%	>90%
Recurrence	Extremely rare (only 5 cases reported)*	Common if underlying HIV is not treated *	Seldom survive to have recurrence

** Ref: Calleros[18], Chidzonga[23]; † Ref: Ghosal[20]*

skin[27] and enzymes such as phosphatase-B that causes bone resorption.[24] *FN* provides growth factor necessary for *Prevotella intermedius* which in turn supports FN by secreting enzymes like lipase, di-peptidyl-peptidases and cysteine proteases that causes lipolysis and destruction of IgG.[28-31] For this reason, these two organisms are sometimes referred to as the 'trigger organisms' of noma.[2,24,32,33] Bolivar identified *Prevotella* and *Peptostreptococus* as the sinister trigger.[34]

Acute necrotizing gingivitis (ANG) is generally acknowledged to be the precursor lesion of noma.[35,33] Marginal necrosis of the interdental papilla serves as the entry point of invasive bacteria. Spreading infection progressively affects tooth sockets (*necrotizing periodontitis*), oral mucosa (*necrotizing stomatitis*), soft tissues of cheek and lips (*necrotizing fasciitis*) and bones (*osteo-myonecrosis*) in that order, thereby establishing the classical noma.[25,36] However, noma is also known to occur without preceding ANG.[25]

Pathogenesis of noma is fraught with many riddles. Most importantly, microbes isolated from the noma wounds do not satisfy the Koch's postulates.[2,24] For example, those organisms incriminated in the pathogenesis of ANG and noma are also found in healthy children. (Table 2) These organisms, when inoculation in laboratory animals, did not produce a disease similar to noma.[24,37] Noma, unlike other infectious diseases, is not contagious. It seldom occurs in more than one member of the family although all of them live in the same socio-economic condition.[38] Hence, synergistic infection appears to be just a triggering factor, while antecedent cause must be different. Several such risk factors have been identified.

Table 2: Frequency of organisms isolated from the oral cavity of children

Organism	ANG or Noma	MN *sans* ANG or Noma	Healthy children
Prevotella	85%	61%	13%
Fusobacterium	30%	39%	38%
Peptostreptococcus	30%	-	-
Campylobacter	55%	28%	-
Streptococcus	*45%*	83%	100%
Enteric Gram negative bacilli	*35%*	28%	-

Data from Falkler[24]
ANG-Acute necrotizing gingivitis; MN- Malnutrition

RISK FACTORS

Oral Hygiene: Poor oral hygiene is a predisposing factor of gingivitis. ANG is common in those who use native 'chew stick' rather than toothbrush.[38] Shared drinking vessel and water contaminated with the feces of livestock are associated with noma.[38] Interestingly, ANG is also seen in more than 2% of children with good oral hygiene and only a tiny fraction of those with ANG develop noma.[2] Therefore, there must be other factors responsible for the transformation of ANG into noma.

Malnutrition: Protein-energy malnutrition (PEM) is invariably seen in all noma patients.[6,8,39] In a large series of 250 noma patients, malnutrition was severe in 55%, moderate in 28% and absent in 7%.[40] But its cause-effect relation-ship with noma is not certain. Although children of a poor family share the same type of food, it is perplexing as to why only one of them develop noma. It is hypothesized that malnourished children may have altered pattern of oral micro-flora.[41] Gram negative anaerobes and spirochetes are more common in them, while gram positive cocci are common in healthy children.[2] Protein deficiency is known to compromise immunity by causing atrophy of lymphoid tissue, reduction of lymphocyte count, alteration of CD_4:CD_8 ratio, malfunctioning of neutrophils and impaired synthesis of immunoglobulins (especially IgA that protects epithelial surface and IgG that is responsible for opsonization). High cellular turn-over of oral mucosa requires constant protein supply to maintain its integrity. Thus, malnutrition is known to cause oral ulcers which may act as entry point of infection. On the other hand, trismus of noma precludes proper feeding thereby contributing to malnutrition.[39] Contrary to the foregoing description, among 69 children with noma, albumin and globulin levels were found to be almost normal.[42] In fact, Eckstein found malnutrition only in those who came to hospital very later, thus suggesting it to be an effect of noma.[35] Enwonwu *et al*. advanced the concept of malnutrition to antenatal period and they noted that maternal malnutrition, as indicated by intra-uterine growth retardation, predisposes to noma by affect the developing thymus of the fetus.[5,43]

Vitamin and Trace Element Deficiency: PEM is often accompanied by varying degree of vitamin deficiencies.[8] Vitamin A prevents atrophy of lymphoid tissues and maintains the epithelial integrity. Vitamins B6 and E are essential for antibody synthesis and cell mediated immunity. Vitamin C influence immunity by altering plasma cortisol level and by affecting bactericidal ability of macrophages and neutrophils. It also causes bleeding ulcers of the gums, delays wound healing and reduce collagen synthesis. B1 and B12 deficiency cause oral ulcers. Deficiency of iron impairs antigen response of leukocytes, while zinc deficiency causes atrophy of lymphoid tissues, affects phagocytosis and impair tissue repair.[8] A combined deficiency of these vitamins and elements is thought to weaken body defense thereby predisposing to noma.

Stress and Cortisol: Psychosocial stress and PEM are known to increase plasma cortisol levels.[8,44] It is more pronounced with marasmus than with kwashiorkor. High cortisol reduces cell mediated immunity, epithelial cell-turn over and collagen synthesis. It also increases serum hyaluronidase

which facilitates bone resorption in noma. Salivary cortisol which matches elevated serum levels is also a growth factor for *Prevotella*, the trigger organism of noma.[2] In addition to cortisol, other stress related hormones such as catecholamines, glucagon, and cytokines are also elevated in noma. All these chemicals may play a role in gluco-neogenesis by breaking down of muscle protein thereby contributing to malnutrition. Psychological stress explain the high incidence of ANG in the post-World War-II concentration camps of Germany and Japan.[3]

Measles: Noma virtually vanished from Europe with the advent of measles vaccination. During a German epidemic 33 of 133 children with measles developed noma.[2] In fact, '*noma-like post measles ulceration*' is a well known entity.[45, 46] Hence it was proposed that measles may predispose to noma either by causing mucosal ulcerations or by post-measles malnutrition.[2, 40] Measles virus is known to impair immunity by killing activated T-cells thereby reducing the production of interleukin-12, increasing interleukin-6 and diminishing retinol.[5] However, a vast majority of noma patients do not have preceding measles.

Herpes - Cytomegalovirus*:* Nearly 59% of ANG patients were sero-positive of CMV.[47] CMV is known to alter immunity by destruction of neutrophils. It also increases production of interleukin-1β which promotes bone resorption in noma. *Herpesvirales* infections such as *Herpes simplex,* CMV and Ebstein-Barr virus are well known to cause micro-ulceration of oral mucosa which could be the entry point of noma-triggering organisms.

Human Immunodeficiency Virus (HIV): Spontaneous noma-like lesion is seen in African monkeys with HIV infection.[4] In a Zimbabwean study[48] all the 27 children with noma were HIV-positive, while in Nigeria none had HIV.[25] The incidence of noma among HIV patients is variously reported as 6-31%.[18] A systematic review identified 24 publications reporting 133 HIV patients with noma.[18] But this is a miniscule proportion of the annual incidence of noma.[18] Therefore, HIV infection appears to be coincidental although immunosuppression of HIV may contribute to noma or noma-like disease. Coexistent HIV increases the risk of noma recurrence [48] and mortality.

Malaria: Noma is common in endemic areas of malaria. Eckstein hypothesized that plasmodium infection may contribute to the onset of noma by affecting reticuloendothelial system, especially the spleen.[35] However, Tempest disagreed with this hypothesis and he found no proof of this association.[40]

Debilitating Illnesses: Several infectious diseases such as typhoid fever, typhus fever, tuberculosis, Leishmaniasis, diphtheria, pneumonia, diarrhea, small pox and chicken pox have been incriminated in the pathogenesis of noma.[2] The exact mechanism is not known. Probably, they predispose to noma by causing malnutrition, poor oral hygiene or mucosal ulcerations. The circular shape of noma gangrene prompted an interesting hypothesis that it could be a form of pressure necrosis of cheek, as debilitated children lie down in bed continuously for several days in lateral position.[4] Debilitating illness are also thought to tilt the balance between pro-inflammatory and anti-inflammatory cytokines, thus facilitating invasive infection.[44]

Malignancy: Noma has been reported to complicate a variety of immunosuppressive disorders such as acute myeloid leukemia,[49, 50] cyclic neutropenia, Down syndrome[51] and Burkitt lymphoma.[52] Immunosupression of chemotherapy drugs is said to cause noma-like disease in adults.[53]

From the foregoing it is possible to hypothesize that noma could be the end result of 3 concomitant factors which, perhaps, form an etiological triangle. (Fig. 1) They are: synergistic

infection of FN and *Prevotella,* impaired immunity and mucosal breech. This is supported by animal experiments in which rats inoculated with bacterial isolates never developed oral mucosal necrosis unless subjected to corticosteroid injections and mechanical trauma of gums.[2,54]

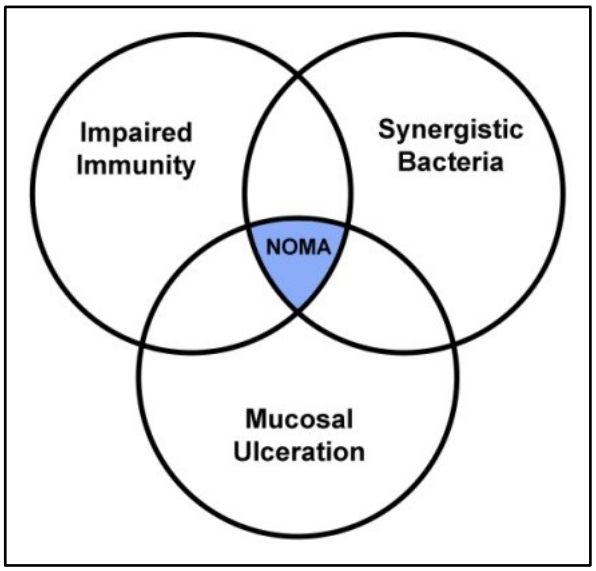

Fig 1. ***Etiological factors of the noma triangle***

DEMOGRAPHY

Exact incidence of noma is not known because many patients with acute disease die in remote villages without getting medical attention.[2,4,55,56] In Nigeria prevalence of ANG is 0.3% and that of noma is 6.4 per 1000 children.[57] Others found 1.6 cases of noma per million population in Nigeria[58] and 4.2 cases per million in Senegal.[59] More interestingly, no new cases of advanced noma were found in the Nigerian survey of 2018.[60] Community prevalence was found to be 3300 per million population in Nigeria.[3] Contrary to these accounts, in 1998, WHO estimated worldwide annual incidence of noma as 140,000 with a prevalence of 770,000 patients.[3,5] This appears to be an overestimate with unknown methodology of calculation [3, 60] A more realistic figure could be 25000 to 40000 new cases per year.[18,61]

Geographic and chronological variation in the incidence of noma is well known. (Fig. 2) In the 1800's noma was frequently reported from Europe and India, in 1900's from Africa and North America and in 2000's from Africa, South America and Asia.[3] Incidence of noma surges following an epidemic of measles.[2] Even within a continent incidence of noma varies from country to country and between rural and urban communities of the same country. The region extending between Senegal and Ethiopia (including Burkina Faso, Mali, Niger and Nigeria), owing to very high incidence of noma, is sometimes referred to as the 'noma belt'.[3, 5, 62] (Fig.2) Incidence of ANG, the precursor of noma, varies between 1-7% in different population.[37, 63]

Several social factors associated with dietary deprivation have been identified as risk factors of noma. They include children raised by grand-mothers,[64] early weaning from breast-milk, absence of chicken at home[2] and families with more than 7 children.[5] *Fusobacterium* is a commensal in the gut of livestock. Feco-oral contamination of animal faeces explains as to why noma is common in dwellers of thatched house, rural communities with unsafe drinking water, barefoot walkers, shared drinking vessels, finger feeding and sleeping on the floor.[38]

Although no age is immune to noma, 80% of patients belong to the age group of 1-6 years.[40] It is seldom seen in edentulous infants and breast feeding babies.[40] William Osler reported high female ratio (M:F - 1:3) from Madagascar, while Durand noted male predominance.[40] Most of the modern series [5, 40] do not find any significant sex predilection. A mild female preponderance may be due to gender discrimination in feeding pattern.

Noma is common in summer months (January-May) when drought and malnutrition are at peak.[5,40] A second smaller peak is seen during winter (July-August) probably due to associated water contamination and malarial epidemics. However some authors deny seasonal variation in the incidence of noma.[40]

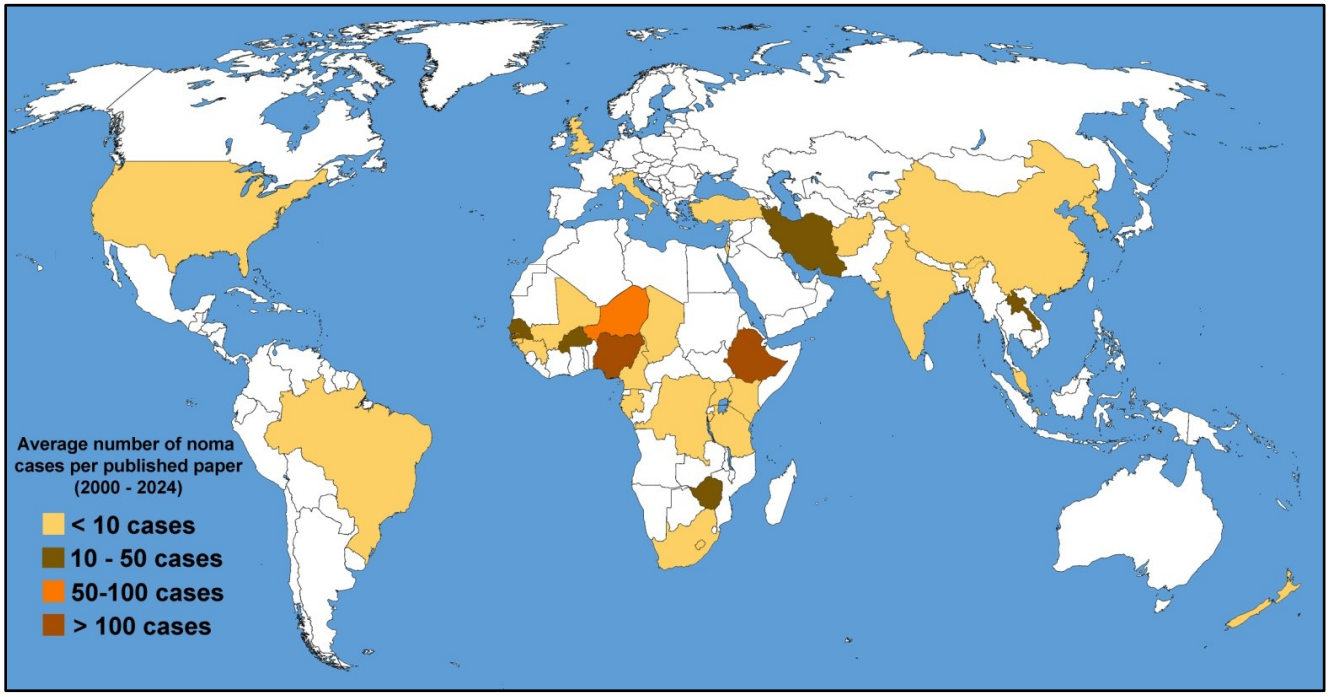

Fig 2. ***Geographic distribution of noma.***

(In the absence of reliable country-wise statistics, the map is constructed based on the number of cases reported from each country as per PubMed records between 2000 and 2024. About 11583 noma patients have been reported in 123 publications in PubMed indexed journals. Country-wise 'case frequency index' was calculated by dividing the number of noma patients reported from a particular country by the number of publications from that geographic location)

PATHOLOGY

Noma usually progresses through a well defined sequence of ANG, necrotizing periodontitis, necrotizing stomatitis, necrotizing fasciitis and osteo-myonecrosis.[36] However, de novo lesions are not unknown.[25]

Noma usually starts at the alveolar margin of pre-molars and mandibular incisors. It is usually uni-lateral; but rarely bilateral. Factors that determine the extent of tissue damage is not known.[25] Although the area of necrosis is not matching with any known pattern of arterial anatomy, noma could still be a vascular phenomenon involving small vessels, capillaries or venous infarction. The necrosed area is in the form of a cone with its apex at skin surface and base at deeper tissues - hence the old name *'cone gangreneux'*.[2] Masticatory muscles and tongue muscles are rarely affected by necrosis but the former are frequently involved in post-gangrene fibrosis.[4]

In very aggressive for of noma, complete destruction of both maxilla and/or mandible occurs.[65] Destruction of bone is perhaps unique of noma differentiating it from other necrotizing fasciitis. Factors incriminated in bone resorption include phosphatase-B secreted by FN, inter-leukin-1β produced by cytomegalovirus and increased serum hyaluronidase in noma. Further, osteonecrosis could also be a vascular event. Involvement of tooth socket may also facilitate onset of early osteomyelitis and bone necrosis.[35] Matrix metalloproteinase an enzyme that is activated by pro-inflammatory cytokines may also contribute to bone resorption.[44,66] Unfortunately, noma affected tissues have not been subjected to proper histopathological studies.[25]

Box 2: Organisms isolated from noma

Gram positive bacilli
Actinomyces israelii [b]
Bacillus cereus [a]
Clostridium beijerinckii [b]
Cutibacterium acnes [1] [b]
Limosilactobacillus fermentum [2] [c]
Lysinibacillus fusiformis [8] [a]
Trueperella pyogenes [3] [c]
Gram negative bacilli
Achromobacter xylosoxidans [a]
Aggregatibacter actinomycetem comitans [c]
Bacteroides fragilis [b]
Brevundimonas diminuta [a]
Brucella anthropi [4] [a]
Cetobacterium [c]
Campylobacter rectus [c]
Capnocytophaga gingivalis [c]
Escherichia coli [c]
Fusobacterium necrophorum * [b]
Fusobacterium nucleatum * [b]
Klebsiella pneumoniae [c]
Mesorhizobium loti [b]
Porphyromonas gingivalis [5] [b]
Prevotella intermedia [6]* [b]
Prevotella melaninogenica [6]* [b]
Prevotella nigrescens * [b]
Pseudomonas aeruginosa * [c]
Selenomonas sp [b]
Stenotrophomonas maltophilia [a]
Tannerella forsythia [b]
Gram positive cocci
Peptostreptococcus micros * [b]
Staphylococcus aureus * [c]
Staphylococcus epidermidis [c]
Streptococcus sp * [c]
Gram negative cocci
Veillonella parvula [b]
Spirochete
Treponema vincentii [7] [c]
Treponema denticola [b]

Former genus name: [1]Propionibacterium, [2]Lactobacillus, [3]Corynebacterium, [4]Ochrobactrum, [5,6]Bacteroides, [7]Borrelia, [8]Bacillus

[a]Aerobe, [b]Anaerobe, [c]Facultative aerobe/anaerobe

** Key pathogenic organisms of noma*

MICROBIOLOGY

Several bacteria have been isolated from noma wounds.(Box-2)[24,67] Molecular studies have shown that only 20-40% of all organisms found in noma lesions could be isolated by conventional bacterial cultures.[2] The principal reason is non availability of anaerobic culture facilities in many resource poor countries where noma is endemic. Paster *et.al.*[68] by molecular studies, isolated 67 different species of microbes from noma wounds. Interestingly, they did not find FN in any of their patients. Large-scale sequencing studies have shown that the identity of isolated organisms are very different from that is reported by conventional techniques.[2]

Bacterial isolates of noma can be classified into primary (causative microbes) and secondary infections (opportunistic commensals invading dead tissue).[8] Clear distinction between the two is often not possible and both of them may contribute to mortality. To exclude secondary pathogens, Eckstein suggested culturing of needle-puncture specimen from tissue adjacent to the margin of necrosis rather than culturing the swab specimens from the gangrenous part.[35] The role of spirochetes in noma is not clear.[24,37,67]

CLASSIFICATION

Noma has two distinct classifications systems: (a) Staging of pathogenesis and (b) Grading of disease severity and extent of tissue damage. Staging is useful in formulating prophylactic and therapeutic strategies, while grading is useful in planning rehabilitative surgery.

Noma Staging

World Health Organisation (W.H.O) originally classified noma into five evolving stages and later added a stage-0. (Box-3)[69] Khammissa *et al.*[36] criticized this classification on several grounds. Although ANG is well accepted to be a precursor of noma, including it in the staging system will inappropriately inflate the statistics of noma. Still

worse is the inclusion of simple gingivitis as the stage-0 which would then mean that one third of the world's population to have noma. According to Khammissa *et al*, this is not only nonsensical but also may potentially distract the scientific understanding of noma as a distinct entity. They also pointed out the inappropriateness of including cheek edema as a stage of noma because it is a non-specific feature seen in several inflammatory disorders. Field workers may mistake edema of other causes for that of noma and vice versa.

Khammissa simplified WHO staging by combining stages 0-1 as 'Noma precursors', stages 2-3 as '*acute noma'* and stages 4-5 as '*arrested noma'*.[36] WHO staging is not without merits, especially in the context of preventive strategies. The statistical and disease-insight concerns of Khammissa *et al*. can be resolved by analyzing stage-specific figures.

From microbiological point-of-view, Falkler[5,8,24] classified noma in to 3 periods: Staging period, Triggering period and Invasive destruction period.

Box 3: Noma staging systems

WHO Staging

Stage 0: Simple gingivitis
Stage 1: Acute Necrotizing Gingivitis
Stage 2: Oedema
Stage 3: Gangrene
Stage 4: Scarring
Stage 5: Sequelae

African Staging

Stage 1: Acute noma
Stage 2: Arrested noma

Noma Grading

There are three popular grading systems of noma: Cariou[4, 70] Montandon[71, 72] and Marck.[73] Cariou classified the resultant defect as simple, extensive and complex. (Box 4) Montandon system is simple and has been adopted by WHO. (Fig. 3) Marck classification is also known as Sokoto or NOITULP classification in which each of the letters stand for a particular anatomical structure: N-nose; O-outer cheek; I-inner cheek; T-trismus; U-upper lip; L-lower lip and P- any other particular problem (e.g. loss of bone or eye). Severity of damage in each of these organs is assessed on a 4 point scale: 0-no loss; 1-loss up to one quarter; 2-loss up to half; 3-loss up to three quarters and 4-near total or complete loss of the anatomical structure. Trismus is assessed on a 4 point scale: T_0 - normal, T_1 - less than 4 cm mouth opening, T_2 - less than 3 cm opening, T_3 - less than 2 cm opening and T_4 - complete trismus. Disease severity of a given patient is then represented by the alphanumeric combination of organ code and severity code (e.g. N_1 O_4 I_4 T_3 U_2 L_1 P-loss of right eye). Obviously, Marck system, albeit accurate, is too complex to follow in clinical practice.

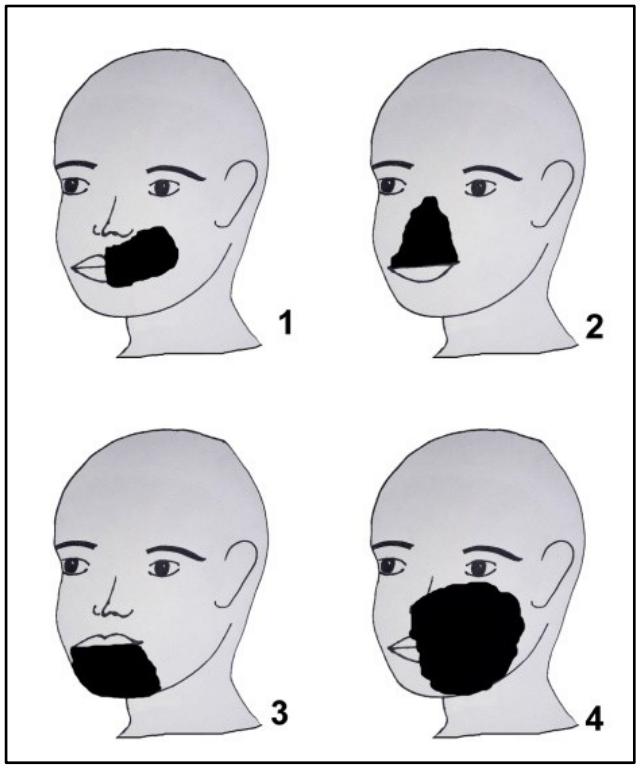

Fig 3. *Montandon (W.H.O) classification*[71]
Type-1 - Defect of lip and cheek involving commissure; Type-2 - Defect of upper lip, maxilla and nose; Type 3 - Defect of lower lip and mandible; Type 4 - Extensive defect of both upper and lower half of face (Art work: Ms. R. Srinidhi)

Box 4: Cariou classification of noma defect[70]

1. **Simple**
   a. Cheek perforation
   b. Commissural destruction
   c. Superior commissuro-labial mutilation
   d. Inferior commissuro-labial mutilation
   e. Supero-medial labial mutilation
   f. Infero-medial labial mutilation
2. **Extensive**
   a. Jugomasseteric mutilation
   b. Labio-mental amputation
   c. Labio-nasal amputation
   d. Labio-maxillo-septo-columellar amputation
3. **Complex**
   a. Lateral hemi-facial lesion
   b. Labio-palatal amputation
   c. Labio-genio-mandibular mutilation

CLINICAL FEATURES

Clinical features of noma differ according to the pathogenic stage of the disease (Table 3 and Fig 4-6).

Fig 4. Stage-1 Noma

Acute necrotizing gingivitis with loss of interdental papillae of the mandibular incisors. (Reproduced from Khammissa RAG, et al 2022, Tropical Medicine and Health, under Creative Commons Attribution 4.0 International License. Ref. DOI:10.1186/s41182-022-00431-6)

Fig 5. Stage 3 and 4 gangrenous noma

Clinical appearance before (a) and after (b) the separation of necrotic eschar. (Reproduced from Khammissa RAG, et.al. 2022, Tropical Medicine and Health, under Creative Commons Attribution 4.0 International License. Ref. DOI:10.1186/s41182-022-00431-6)

Fig 6. Noma Neonatorum

(Courtesy: Prof. Raveenthiran V. Image not covered under Creative Commons licence. Presented with the permission of parents)

Table 3: Clinical features according to WHO staging of pathogenesis *

WHO Stage	Clinical features
0 (Gingivitis)	Malnutrition, Vitamin deficiency, Anemia, Bleeding, pain, swelling and redness of gums, Listlessness, Apathy
1 (ANG)	Loss of interdental papilla, Halitosis, Excessive salivation, Cervical lymphadenopathy, Pyorrhoea, Fever, Metallic taste
2 (Edema)	Edema of cheek or lips, Acute trismus, Painful mouth opening, Greyish-black ulcers in mucosal aspect of mouth, Leukocytosis
3 (Gangrene)	Black discoloration of perioral skin, Loss or deformity of teeth, Full thickness necrosis of cheek, nose, jaws or lips, Features of systemic sepsis such a fever, tachycardia, lethargy, circulatory shock, tachypnoea, anorexia, fever, dehydration
4 (Scarring)	Established orocutaneous or oronasal fistula, Complete necrosis of cheek, nose, lips and facial bones, Temporo-mandibular ankylosis, Chronic trismus, Feeding difficulties, Malnutrition, impaired articulation of speech
5 (Sequelae)	Regurgitation of feeds, Impaired speech, Cosmetic mutilation of face, Psychological depression, Salivary fistula, Epiphora, Entropion, Exposure keratitis, Corneal ulcers, Loss of lower eyelid, Enophthalmos due to orbital floor necrosis, Deformed permanent teeth, Malocclusion, Anosmia and breathing difficulty due to nasal disfigurement

** Some features of a given stage may overlap with those of preceding and following stages. ANG - Acute necrotizing gingivitis*

DIFFERENTIAL DIAGNOSIS

In pre-gangrene stages constitutional symptoms of noma may be mistaken for febrile illness such as malaria. In *prodromal stages*, noma is indistinguishable from acute herpetic gingivo-stomatitis, acute oral candidiasis and Vincent stomatitis. Facial cellulitis, angioneurotic edema, maxillary rhabdomyosarcoma, Burkitt lymphoma, osteosarcoma and maxillay sinusitis may mimic the *edema stage* of noma. Several necrotic and granulomatous lesions may be mistaken for the mutilating *gangrenous stage* of noma. They include muco-cutaneous Leishmaniasis, chronic lupus erythematosis, syphilis, yaws (gangosa), midline granuloma of Wegener, mucormycosis, oral cancers, lepromatous leprosy, agranulocytic gangrene, clostridium gangrene, lupus vulgaris, ecthyma gangrenosum, Stewart's granuloma, radiation necrosis and sickle cell crisis.[5,6] Grossly deformed face of noma may resemble Romberg disease (progressive facial hemiatrophy) and post-burn contractures. Acute trismus of the noma neonatorum should not be confused with neonatal tetanus.

INVESTIGATIONS

Noma is a clinical diagnosis. However, underlying immuno-suppressive disorders such as HIV, leukemia, anemia and protein malnutrition should be excluded by appropriate lab investigations. Inflammatory markers such as leukocyte count, erythrocyte sedimentation rate, tumor necrotic factor-α, interleukins, and C-reactive protein will be elevated. Although non-specific of noma, they are very useful in monitoring the disease progress during therapy. Very low CD4 count is typical of HIV associated noma.[74] Blood cultures are invaluable in guiding antibiotic treatment. On the other hand, wound swab cultures are not very helpful as they often fail to differentiate primary infection from secondary contaminants. Computed tomography with 3-D reconstruction is essential in planning reconstructive surgery especially when the jaw bones are destroyed.[75]

TREATMENT

Management of Acute Noma

Progression of ANG to gangrenous noma can be arrested by restoring the oral hygiene.[76] This includes using a toothbrush, antiseptic (chlorhexidine) mouthwash and oral antibiotics. Dental scaling, though useful in removing plaques, should be avoided in the presence of active infection, as it may precipitate bacteremia.[22,72] Mobile shaky teeth may be removed. Simultaneously, protein malnutrition, vitamin or trace element deficiencies and anemia should be corrected. Re-feeding syndrome may complicate aggressive nutritional rehabilitation. In moribund patients a brief course of corticosteroids may be helpful.[4] Anecdotal observation suggests that topical phenytoin may be helpful in improving oral health by causing gingival hyperplasia (*Unpublished observation of the senior author - VR)*.

Management of acute noma necessitates hospitalization. In addition to observation of oral hygiene, prompt correction of dehydration, electrolyte imbalance and intravenous administration of broad-spectrum antibiotics are indicated.[25] Severe trismus may necessitate nasogastric tube feeding or supplemental parenteral nutrition. Daily wound cleaning with hydrogen peroxide, povidone-iodine or Eusol is recommended.

Antibiotics are often chosen empirically and rarely on the basis of culture guidance. Chosen combination of antibiotics should cover both aerobic and anaerobic bacteria.[2,22,25,76] Typically the triple combination of a penicillin with clavulanic acid or a third-generation cephalosporin *plus* an aminoglycoside *plus* metronidazole is used. Ciprofloxacin, fluconazole, linezolid and streptomycin are less commonly used. Although clindamycin is very effective against anaerobes, metronidazole is preferred over it to avoid pseudomembranous colitis. Underlying comorbidities such as HIV should also be simultaneously treated by antiretroviral drugs.

Role of Surgery in Acute Phase

Aggressive debridement of gangrenous tissue is generally not advised in the acute stage as it may precipitate fatal bacteremia. Spontaneous, slow separation of eschar is less mutilating than active surgical excision.[4] Occasionally, gentle surgical debridement of eschar and sequestrectomy may be required.[77] After eschar removal, the wound is best allowed to stabilize by secondary healing. No attempt of reconstruction should be done within one year of recovery from acute illness.[25,77] During this waiting period, attention should be diverted towards improving nutrition, treating concomitant immuno-deficiency disorders and preventing temporo-mandibular joint (TMJ) ankylosis. Splinting of the jaws, mouth opening devices and physiotherapy may minimize chronic trismus.[78] Muscle interposition after osteotomy of the mandibular condyloid process may avoid recurrent ankylosis. Psychological rehabilitation should also be started simultaneously.[79]

Anesthetic Considerations

Severe ankylosis of TMJ and poor mouth opening often cause serious problem with endotracheal intubation during anesthesia.[81] This problem is best overcome by fiberoptic bronchoscope aided intubation. Alternatively trans-tracheal induction of anaesthesia, blind nasal intubation, preliminary tracheostomy, excision of coronoid process or osteotomy of the mandibular angle under local anesthesia through a submandibular approach can be done.[4]

Water's technique of endotracheal intubation is an ingenuous method of bypassing severe TMJ ankylosis.[40, 82] In this technique a Tuohy needle with a Huber point is inserted into the trachea through the cricothyroid membrane under local anesthesia. An epidural catheter is then threaded through the needle. The bevel of the needle tip is positioned facing upwards so that the catheter is directed superiorly to pass through the vocal cords into the nasopharynx. The coiled catheter from the nasopharynx is then retrieved by passing

another hooked catheter or endoscopic grasping forceps through nostrils. Keeping the tightly held epidural catheter as guide endotracheal tube is slid into trachea and the guiding catheter removed.

Reconstructive Surgery

Surgical reconstruction of noma defect offers a fertile field for imaginative surgeons. Noma rehabilitation is a team work involving pediatric surgeon, plastic surgeon, maxillofacial surgeon, pedodontist, clinical child psychologist, physio-therapist and community nurse. All the known principles of plastic surgery have to be employed on case-to-case basis and individual tailoring of treatment is the key principle. The order of complex, staged reconstructions include relief of trismus, functional restoration of the oral cavity, reconstruction of bone defect and finally cosmetic fine-tuning.[4,40,77,80]

Lip defects are repaired by Abbe-Eslander flaps or Gillies fan flap.[83] Pedicled forehead flaps and condro-cutaneous flaps from the ear are popular in reconstructing the nose. The full-thickness cheek defects are repaired by a variety of techniques including Dijkstra's platysma flap, Bakamjian's medial based deltopectoral flap,[84] prefabricated scapular flap,[85] supraclavicular flap,[86] lattisimus dorsi myocutaneous flap, tubularized distal pedicle (waltzing) flap[87] and free microvascular flaps.[88] Lattisimus dorsi flaps are preferred over pectoral flaps in females for obvious cosmetic reason. Palatal defects are repaired using galeal or temporal flaps.[4, 40] If simultaneous reconstruction of mandible along with cheek defect is required, forearm osteo-myocutaneous flaps with radial bone is preferred.[89] Lower eyelid contractures may be corrected by Z-plasty or by Gillies bipedicled upper eyelid flap. Severe local scarring often precludes the use of skin expanders in reconstruction.

COMPLICATIONS

Distorted facial growth is a common complication of noma. Metastatic gangrene of the neck, scalp, perineum and genitalia are not uncommon.[4] Malignant transformation of the noma ulcer,[90] maggot infestation,[91] and recurrent parotitis due to distorted salivary duct[92] have rarely been reported.

Following complex facial reconstruction, recurrent trismus due to TMJ ankylosis or scar hypertrophy is known to occur. Facial disfigurement, social isolation and local taboos cause severe psycho-logical depression in one third of the noma survivors.[79,93] Surgery of noma is fraught with all the known complications of reconstructive surgeries such as wound infection, flap necrosis and scar contracture.[94]

PREVENTION

Health education of people, awareness creation among health care professionals, easy access to health care facility, encouraging the usage of toothbrush, mitigation of extreme poverty and malnutrition, segregation of livestock from human inhabitation, provision of safe drinking water and promotion of breast feeding are of paramount importance in preventing noma.[5] Vaccination against measles may reduce the vulnerability of the at-risk population. Noma is non-contagious and hence isolation of affected patients is unnecessary.[3]

OUTCOME

Untreated noma is invariably fatal (80-90%). Most of the children die due to secondary infection, septicemia, malnutrition, aspiration pneumonia and mental depression.[2] Coexistence of HIV increases mortality and it is a risk factor of recurrent noma.[18] With proper treatment, the current mortality of noma is less than 10%.[3]

Recurrent noma is extremely rare.[18,40,77] In a series of 580 children with noma from Zinder,

there were no recurrences.[3] Those who survive acute noma usually live long into adult life, albeit it with severe morbidity.

CONCLUSION

Noma is a preventable opportunistic infection. Maintenance of personal oral hygiene will largely avoid noma. However, other risk factors such as alleviation of poverty and malnutrition, provision of safe drinking water and infrastructural facility to do complex reconstructive surgeries are to be addressed by governments and political will. Therefore, WHO rightly mentioned, *"Noma is more than a disease"*.

Epilogue

Among the published manuscripts on Noma 72% are case reports/series, 10% are retrospective studies, 11% are cross sectional studies, 5% are prospective case control studies.(Farley 2021) [3] *Randomized trials and meta-analysis are not available. Available systematic reviews are descriptive rather than analytical. Hence, opinions expressed herein are of level-4 evidence*.

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Diagnostic Image

Split Notochord Syndrome

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Fig 1. Plain radiograph showing split notochord at the sacrum

Split notochord syndrome is a rare malformation, with only fewer than 35 cases reported in the world literature until 2017.[1] The child depicted herein presented soon after birth with a lesion resembling myelomeningocele in the sacral region. It was a soft, pink, fleshy mass. There was no neurological deficit of lower limbs. During surgical operation, a communication between the dural sac and the rectum was discovered. It was securely closed with 4-0 silk sutures. Dura was closed in routine manner. Patient made uneventful recovery and he continues to remain well.

Split notochord malformation has the following components: (a) A wide defect in the vertebral column; (b) Communication between the neural axis and intestinal tract; and (c) Open neural tube defect externally. Many variations have been described. Neuraxis may communicate with the ileum, cecum, colon or the rectum (as in this patient). There may be associated meningocele, neurenteric cysts or sacral agenesis.

The yolk sac and the amniotic cavity communicate with each other during the third week of gestation. Persistence of this communication can lead to formation of neurenteric canal.

Management of split notochord should be tailored individually according to the nature of anatomical variations. Considering its rarity, wide variations in presentation and heterogenous pathological anatomy, management guidelines do not exist. Surgeon should aim to achieve complete separation of the neural and enteric systems without causing operative neurological damage. Long-term follow-up is essential to detect late sequelae and treat complications.

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Clinical Update

On the Role of Prophylactic Gonadectomy in Turner Syndrome

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Keywords
Germ cell tumor
Gonadoblastoma
Karyotype
Testis specific protein Y
Y-chromosome

Abbreviations
GB - Gonadoblastoma
GCNIS - Germ cell neoplasia in situ
GCT - Germ cell tumors
PCR - Polymerase chain reaction
TS - Turner syndrome
TSPY - Testis specific protein-Y

Abstract

Turner Syndrome (TS) is a chromosomal disorder with a karyotype of 45X. However, by conventional karyotyping, Y-chromosomal material is known to occur in up to 5-12% of TS patients; but it escalates to 60% when molecular diagnostic tools are used. The most commonly investigated genes were SRY, DYZ3, ZFY, and TSPY. Identifying the Y-chromatin in TS is important as it is associated with the risk of developing germ cell tumors, especially gonadoblastoma (GB). The prevalence of GB in TS is variously reported as ranging from 0 to 33%. As imaging studies cannot conclusively exclude malignancy of abdominal gonads, prophylactic gonadectomy is recommended in TS patients with Y-chromatin fragment. However, the optimal time of prophylactic gonadectomy is not known. The International Turner Syndrome Consensus Group recommends gonadectomy at the time of diagnosis, regardless of age while recent evidences suggest a more flexible approach, especially at puberty.

INTRODUCTION

Turner Syndrome (TS) is a genetic disease characterized by short stature, sexual infantilism, gonadal dysgenesis, and typical dysmorphic features. Its prevalence is approximately 1 in 2000–2500 live female births.[1] About 40%–60% of TS patients have a 45X karyotype, and the others have structural abnormalities of the X-chromosome or various kinds of mosaicism. In addition, a cell line containing a Y-chromosomal segment is present in up to 60% of TS patients.[2] Identifying Y-chromatin in TS individuals is essential as it is associated with the risk of developing germ cell tumors (GCT). Gonadectomy is generally recommended in TS females with Y-chromosome-derived material to prevent the development of GCT, especially gonadoblastoma (GB).

PREVALENCE OF Y-CHROMOSOME MATERIAL AMONG TURNER SYNDROME PATIENTS

Karyotyping identifies a normal or structurally aberrant Y-chromosome in 5–12% of the TS patients.[3] However, molecular studies such as polymerase chain reaction (PCR) and fluorescence in situ hybridization (FISH) can detect concealed Y chromosome material in TS patients, consequently escalating the Y-chromosome sequence detection frequency to approximately 60%.[3-4] The most commonly investigated genes were SRY, DYZ3,

ZFY, and TSPY.[1,3-4] To detect individuals at high risk of GCT, the TS Consensus Study Group recommended testing for Y-chromosome material only in TS patients who present with a marker chromosome or virilization.[5] However, many recent studies have reported molecular identification of Y-chromosome material in TS patients even in the absence of virilization or a marker chromosome. This finding suggested the necessity of screening for cryptic Y-chromosome in all TS patients.

PATHOGENESIS OF GERM CELL TUMORS

The biology of the development of GCT is not completely known. The peri-centromeric gonadoblastoma region of the Y-chromosome (GBY locus) is deemed to contain the presumed GB gene, for example, the testis-specific protein-Y (TSPY) gene.[6] TSPY behaves as a proto-oncogene in patients with dysgenetic gonads. An interaction of the OCT3/4 gene with TSPY is implicated in the pathogenesis of GB. OCT3/4 is essential to maintaining the pluripotent nature of embryonic stem cells and primordial germ cells.[6] GB, classified as a borderline GCT arising from sex cord and germ cells, is intrinsically linked with gonadal dysgenesis. Y-chromosome-associated GB can transform into invasive dysgerminoma or other malignant GCT, including embryonic carcinoma, teratoma, yolk sac tumor, and choriocarcinoma.

RISK OF DEVELOPMENT OF GERM CELL TUMORS

The risk of developing GB in TS patients with Y-containing material is variously quoted in several studies. As TS patients with multiple Y-variants are grouped under a single cohort in most studies, the risk stratification based on the levels of mosaicism, inclusion or exclusion of TSPY, translocated portions of the Y and other Y chromosome variants is lacking in the literature. In a systematic review by de Marqui *et al.*[4] nine studies reported the prevalence of GB ranging from 0 to 33%. In another review by Pyle *et al.*[7] 26 cases had germ cell neoplasia in situ (GCNIS)/GB (26.2%), and three patients had dysgerminoma (3%) among 99 TS patients with Y-chromosome material included from 7 studies published after 2006. In contrast, a UK cohort study published within the same time frame including 212 patients described five (2.4%) with GCNIS/GB and two (1.0%) with dysgerminoma.[8] A meta-analysis of 11 studies analyzing the association between Y-chromosome material and GB reported five cases of GB in 43 patients (11.6%) with TS-bearing Y-chromosome cell lines.[9]

The diagnosis of GCT primarily depends on pathological assessment following gonadal biopsy or gonadectomy.[3] However, multiple histological sections of the streak gonad are required to demonstrate microscopic GB. Neither ultrasonography nor magnetic resonance imaging can definitively exclude neoplasms of intra-abdominal gonads in TS patients. It may be bilateral in up to 10% of cases.[10] Since not all TS individuals with Y cell lines develop tumors, other unrecognized factors must also be present predisposing to tumor formation. Cools et.al.[11] reported a correlation between external genital phenotype as assessed by external masculinization score and presence of GCNIS/GB. However, this finding could not be replicated in other studies.[11] The clinical features of virilization may not be present in all TS subjects with GCT. Similarly, a Y-positive TS will have a higher GCT risk with abdominal gonads as compared to scrotal gonads, and the risk increases with advancing age.[7] Furthermore, the risk of GB development among patients with cryptic Y-chromosome material is similar to that of individuals possessing overt abnormalities of the Y-chromosome.[12] However, the only established risk factors for malignant transformation are older age and intra-abdominal location.[13]

ROLE OF GONADECTOMY

Coyle *et al.*[6] observed GB in four of 14 patients, of whom only one was older than six years and the youngest was just five months. Matsumoto *et al.*[14] reported a series that included seven females with

TS having Y-chromosome material, aged between 2 and 11 years. Pathologic examination revealed gonadal tumors in four subjects, the youngest of which was 2 years old. The fact that GB is thought to occur most commonly in the second decade of life may be a result of late diagnosis of TS in other series, leading to later prophylactic gonadectomy. If gonadal biopsy is done before determining the appropriateness of gonadectomy, histological examination of the biopsy specimen for TSPY and OCT3/4 expression has been recommended to improve the detection rate of GB and germ cell malignancy.[6]

Although the risk of GB in individuals with a Y-chromosome is not precisely quantified, gonadectomy for individuals with a fragment of Y-chromosome should continue to be mandatory as recommended by the recent guideline.[15] The International Turner Syndrome Consensus Group recommends that female individuals with TS and Y-chromosome material should undergo gonadectomy at the time of diagnosis, regardless of age.[15] However, others suggest gonadectomy should be considered at puberty.[7] This view is supported by the data that 42% of girls with TS with Y-chromosome material attained puberty spontaneously and 11% achieved spontaneous menarche indicative of gonadal function.[13] Recent developments have also occurred in the medical and surgical management of TS patients with GCT. Therefore, gonadectomy is no longer considered the default treatment approach for TS subjects with Y-chromosome material at the first visit. The timing of such surgery and risk of malignancy can be discussed with parents during counseling.

CONCLUSION

The prevalence of Y-chromosome material among TS subjects should be meticulously evaluated by PCR as it is inexpensive, sensitive, rapid, and allows the tracking of various sequences of Y-chromosome simultaneously. In those patients with Y-positive sequences, prophylactic gonadectomy is recommended for prevention of GB. However, the age at which the gonadectomy to be offered is inconclusive. Given the benign nature of GB there seems to be considerable advantage in doing gonadectomy at puberty.

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How I Do It

Removal of Double-J Stent in Children without Anesthesia or Cystoscopy: A Useful Technique

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Keywords
Double-J stent removal
Urological surgery
Cystoscopy

Abbreviations
DJ Stent - Double J Stent

Abstract

Background: *Double-J (DJ) stents are commonly used in pediatric urology, especially during surgical procedures like ureteric re-implantation, pyeloplasty, pyelolithotomy and percutaneous nephrolithotomy. Postoperatively removal of the DJ stents requires cystoscopy under anesthesia. We present our experience with a novel technique of DJ stent removal without the need of cystoscopy.*

Technique: *DJ stent was deployed with a 4-0 silk thread tied to its bladder end. In post-operative period the thread is spontaneously passed out of external urethral meatus during per-urethral voiding. On the postoperative day-7, the string was simply pulled at bed side to remove the stent.*

Result: *We used this technique successfully in 7 consecutive patients. There were no complications in this series.*

Conclusion: *Ureteral stent removal using an extraction suture is technically feasible. It is beneficial because it obviates an invasive cystoscopy, thus decreasing the cost of healthcare in limited-resource settings.*

INTRODUCTION

Double-J (DJ) stents are commonly used in pediatric urology, especially after surgical procedures such as ureteric re-implantation, pyeloplasty, percutaneous nephrolithotomy and pyelolithotomy. Post-operatively, stent removal requires cystoscopy under general anesthesia. Re-admission to hospital, considerable operating-room time, need of general anesthesia, special equipments and invasiveness of the procedure are the disadvantages of this approach.[1, 2]

Earlier, pediatric surgeons were using small-sized polythene tubes for stenting ureteral anastomosis after re-implantation or pyeloplasty. One end of these tubes was exteriorized through a small stab wound. These stents were left in situ for a week and later removed by pulling the outer end of the tube. This is difficult to handle as there is chance of unintentional, accidental, premature removal. We describe a technique of DJ stent removal by using an extraction string which obviates the need for cystoscopy.

THE TECHNIQUE

DJ stent is deployed with a short length (10-12 cm) of 4-0 silk tied to its distal (bladder) end. In the post-operative period, under the influence of fluid current of voiding, the thread spontaneously extrudes out of urethral meatus within 3 to 4 days. If required the tread is strapped to penile shaft to avoid accidental pulling. On the seventh or eighth post-operative day, both the thread and stent are painlessly pulled out at bed side.

Fig 1. The extraction suture technique
Silk thread attached to double-J stent is seen emerging through the external urethral meatus

CLINICAL EXPERIENCE

This is a prospective study conducted at the Rural Medical College, Loni, Maharashtra. After an open surgical procedure on the pelvi-ureteric junction (PUJ) or vesico-ureteric Junction (VUJ), DJ stent was inserted if it was considered necessary. The described technique was used in 7 patients with 10 DJ stents. (Table 1) The age group of patients was 1 to 7 years. Only those urological procedures which required short term DJ stenting were included in the study. The DJ stent was removed by string extraction technique on the post-operative day-7 in all the 7 patients. Anesthesia, sedation or cystoscopy was not required in any of them. The stent could be easily removed in all of them without any complication.

Table 1: Clinical details of patients who underwent double-J stenting

Pt. No	Age	Sex	Side	Indication
1	2 yr	M	Unilateral	Ureteric reimplantation
2	3 yr	F	Unilateral	Nephrolithotomy
3	5 yr	M	Bilateral	Pyeloplasty
4	6 yr	M	Bilateral	Pyeloplasty
5	1 yr	F	Unilateral	Pyeloplasty
6	2 yr	M	Unilateral	Ureteric reimplantation
7	7 yr	M	Bilateral	Pyeloplasty

DISCUSSION

DJ stents have a prominent role in pediatric urology, especially after ureteric operations.[2] Our technique has several advantages. It does not require cystoscopy or general anesthesia. Readmission to hospital and the cost associated with it are avoided.[3] Special equipments like cystoscopic grasper are not needed. It is a safe and effective method without any complications.[4] Patients with cystinuria can remove their stents at home.[5]

DJ stents are usually left in situ for a week. However, in special situations, some surgeons prefer to leave them in situ longer period. In such cases, not infrequently, patients and surgeons forget to remove the DJ stents.[1] This risk of long term retention of a DJ stent is eliminated in our technique since the extraction string is externally visible.

Small sample size is a limitation of our study. This technique requires patient compliance. The care-givers have to be counseled about the procedure and purpose of the string, lest they may mistakenly pull it out.

CONCLUSION

The string technique of DJ stent removal is a safe and economical alternative to the conventional

technique of cystoscopic removal which is invasive and expensive.

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Clinical Study

A study of retroperitoneal core needle biopsy in the diagnosis of suspected malignant renal tumors

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Keywords
Clear cell sarcoma of kidney
Core needle biopsy
Kidney tumors in children
Malignant rhabdoid tumor
Non-Wilms renal tumor
Renal tumor
Wilms tumor

Abbreviations
CCSK - Clear cell sarcoma of kidney
CECT - Contrast enhanced computed tomography
CNB - Core needle biopsy
COG - Children's oncology group
HPE - Histopathological examination
MRTK - Malignant rhabdoid tumor of kidney
NACT - Neo-adjuvant chemotherapy
NWRT - Non-Wilms renal tumors
NWTSG - National Wilms tumor study group
RTGS - Renal tumor study group
SIOP - Societe Internationale D'Oncologie Pediatrique
USG - Ultrasonography
WT - Wilms tumor

Abstract

Background: *Differentiation of Wilms tumor (WT) from non-Wilms renal tumors (NWRT) is essential as the management philosophies of these two groups of tumors are very different, especially if we believe in the SIOP protocol of administering neo-adjuvant chemotherapy (NACT) in patients with WT. This study examines the role of the preoperative retroperitoneal core needle biopsy (CNB) in the differential diagnosis of suspected malignant renal tumors in children*

Material and methods: *CNB was done in 20 patients (23 renal biopsies as 3 patients had bilateral tumors) with suspected malignant renal tumors. Those diagnosed as WT on CNB received NACT followed by nephrectomy and adjuvant therapy. We studied different subtypes of WT, chemotherapy-induced changes in the resected specimen and the concordance between the histopathology examination (HPE) of pre-operative CNB with that of the excised surgical specimen.*

Results: *Out of the 23 renal biopsies, 21 were diagnosed as WT while the other 2 as malignant rhabdoid tumors of kidney (MRTK). One patient developed abdominal distension and respiratory distress after CNB. On HPE of the excised surgical specimen of those who received NACT for WT (diagnosed on CNB), 19 samples were confirmed as WT, while 2 were diagnosed as clear cell sarcoma of the kidney (CCSK). Of the 2 tumors that were diagnosed as MRTK on CNB, 1 was confirmed as MRTK and the other was diagnosed as CCSK on HPE of the excised specimen. The concordance between the HPE of CNB and the excised surgical specimen was 91%.*

Conclusion: *Pre-therapy CNB is a sensitive tool to differentiate WT from NWRT. The sensitivity, specificity, positive predictive value and negative predictive value of CNB in the diagnosis of renal tumors are 89%, 50%, 89% and 100% respectively.*

INTRODUCTION

Childhood cancers represent a significant health concern worldwide, with malignant renal tumors accounting for approximately 7% of all childhood cancers. Among these, Wilms Tumor (WT) is one of the most common, accounting for about 85% of all pediatric malignant renal tumors.[1] Accurate diagnosis of renal tumors is crucial for selecting the most appropriate treatment and ensuring optimal outcomes for patients. It is not always possible to differentiate between different types of renal tumors by clinical examination and imaging alone.[2] Saula *et al.* noted approximately 13.6% of cases were NWRT in the developing countries, highlighting the importance of preoperative CNB in guiding treatment decisions.[3] Moreover, NWRT are far more commonly seen in Southeast Asian countries including India as compared to the Western world.[3] WT is treated according to the guidelines suggested by the National Wilms tumor study group (NWTSG) - recently renamed as Children's Oncology Group (COG) - or the International Society of Pediatric Oncology (SIOP).[4] NWTSG/COG believes in up-front surgery followed by adjunct therapy, while SIOP believes in neo-adjuvant chemotherapy (NACT) followed by surgery and then adjuvant therapy.

During SIOP trials, it was noted that 5-10% of tumors initially suspected to be WT turned out to be either benign renal tumors or NWRT that do not respond to NACT.[5] COG approach has the advantage of studying tumor histology unaltered by prior chemotherapy and hence it makes subtyping and staging of the tumor easier. Even in COG, NACT is administered to inoperable WT, WT in a solitary kidney, synchronous bilateral WT, tumor thrombus in inferior vena cava extending above the level of the hepatic veins and the tumor involving contiguous vital structure.

In India, pre-treatment CNB is not a popular practice, unlike in the United Kingdom where the Children's Cancer Study Group (UKCCSG) used to advocate preoperative CNB through the retroperitoneal route for all suspected WT to ascertain the histopathological diagnosis before administering neo-adjuvant chemotherapy.[5] As the management philosophy of WT and NWRT is very different, we intended to assess the role of the pre-therapy retroperitoneal CNB in the differential diagnosis of pediatric renal.

MATERIALS AND METHODS

After obtaining the approval of the institute ethical committee (LHMC/IEC/2021/03/88), a prospective descriptive study of 20 patients with suspected malignant renal tumors was done during the period of July 2021 to November 2022. Patients with recurrent disease and tumor rupture or hemorrhage necessitating emergency surgery were excluded. All the patients had routine biochemical investigations, ultrasonography (USG) abdomen with Doppler and contrast enhanced computed tomographic scan (CECT) of the chest and abdomen. (Fig. 1)

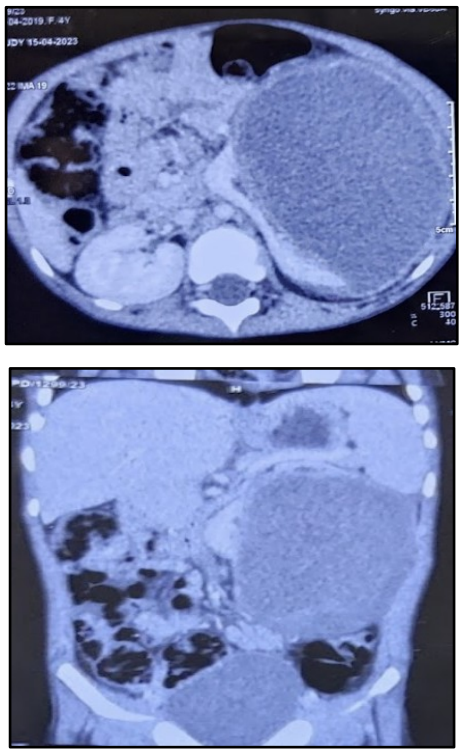

Fig 1. Left renal Tumor

Contrast Enhanced Computed Tomography (CECT) showing the left renal mass in axial (upper panel) and coronal (lower panel) section.

Fig. 2: USG guided core-needle biopsy procedure.

All but one patient underwent USG-guided CNB through the retro-peritoneal route after administering adequate sedation and local anesthesia; one patient erroneously underwent transperitoneal CNB.(Fig.2) At least 2-3 cores were taken and sent for HPE. Adverse events, either during or after the procedure, were recorded. HPE was reported within approximately 5-6 days. For patients with bilateral renal tumors, both tumors were biopsied as discordant pathologies are known to occur. Those who were diagnosed as WT were given NACT as per the Umbrella protocol of SIOP-RTSG[18] and those diagnosed as MRTK underwent upfront surgery. In CNB diagnosed WT, a re-evaluation was done by CECT of the abdomen before nephrectomy to assess the reduction in tumor volume due to NACT.

Written consent for surgery was obtained from caregivers. All patients underwent tumor excision by a single senior surgeon (YKS). Sixteen patients underwent total nephro-ureterectomy; these include 2 children diagnosed to have MRTK on CNB. Four patients underwent nephron-sparing surgery; 3 of them had bilateral WT. Intra-operative and post-operative adverse events, if any, were recorded. The excised surgical specimen was sent for HPE (Fig. 3) to study tumor staging and risk-stratification. Concordance between the pre-therapy CNB report and that of post-nephrectomy HPE was recorded. NACT-induced changes in histopathology were noted. In the case of bilateral tumors, adjuvant therapy was based on the tumor having the higher risk stratification.

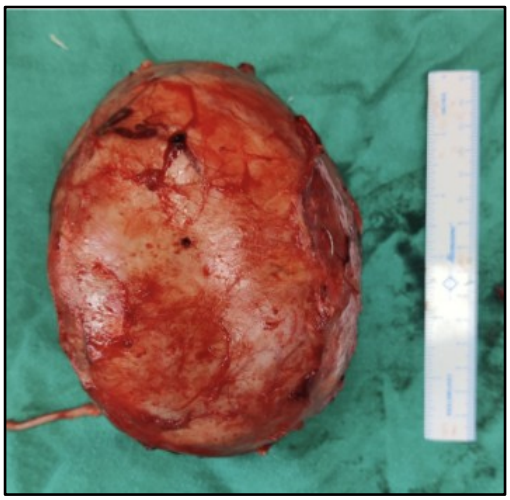

Fig. 4: Left nephro-ureterectomy specimen

RESULTS

CNB was performed for 23 renal tumors in 20 patients; 3 had bilateral renal tumors. In one patient, the CNB was done transperitoneally by mistake. She developed abdominal distension, respiratory distress and a fall in hemoglobin due to intra-abdominal bleeding. She was upstaged to stage-3 and was treated accordingly. CNB-related hemorrhage, tumor rupture or tumor seeding was not noted in any other patient.

Eighteen patients including 3 having bilateral WT (21 renal CNB) were diagnosed to have WT. They received NACT as per SIOP protocol. The other 2 patients diagnosed as MRTK on CNB underwent upfront nephrectomy.(Table 1). More than half of the WT (57.1%) patients were reported to have biphasic WT on CNB.

The interpretation of monophasic WT with blastemal component alone (n=2) on CNB was easy to interpret. However, interpreting monophasic WT with stromal component (n=2) needed considerable expertise. In 19% (4/21) rhabdomyoblastic changes were seen.

Table 1: Histopathological diagnosis on core-needle biopsy

Renal tumors		n (%)	Blastemal	Epithelial	Stromal	RMB
WT	Monophasic	4 (19%)	2	-	2	-
	Biphasic	12 (57.1%)	8	9	7	3
	Triphasic	5 (23.8%)	5	5	5	1
MRTK		2 (8.6%)	-	-	-	-

MRTK - Malignant rhabdoid tumor of kidney, RMB - Rhabdo-myoblastic change; WT - Wilms tumor

Table 2: Histopathological diagnosis of the nephrectomy specimens

Tumor Histology with risk stratification		*n* (%)	Blastemal	Epithelial	Stromal	RMB
WT (n=19; 82.6%) **IR-17** Mixed type - 9 Stromal type - 4 Regressive type - 3 Epithelial type - 1 **HR** (Blastemal type) **-** 2	Monophasic	1 (5.2%)	-	1	-	-
	Biphasic	5 (26.3%)	-	5	5	2
	Triphasic	13 (68.4%)	13	13	13	3
CCSK (HR)		3 (13%)	-	-	-	-
MRTK (HR)		1 (4.3%)	-	-	-	-

CCSK- Clear cell sarcoma of kidney; HR- High risk; IR- Intermediate risk; MRTK- Malignant rhabdoid tumor of kidney; RMB - Rhabdo-myoblastic change; WT-Wilms tumor.

None of the WT showed anaplasia. The mean reduction of tumour volume after NACT was 41.8 $\pm$ 2%. No change in the tumor volume after NACT was noted in 13/21 (61.9%) tumors. None of the bilateral tumors (n=6) or the 2 tumors that later turned out to be CCSK showed any change in tumor volume. Only 3 patients with unilateral WT on CNB showed considerable (>50%) reduction in tumor volume following NACT. Heterologous elements like cartilage and bone were found in 2 specimens, while rhabdo-myoblastic changes were seen in 5 specimens.

On histopathology of the excised specimens, 17 (73.9%) patients had intermediate-risk tumors and 6 (26%) had high-risk tumors as per working SIOP classification; none was of 'low-risk' category (completely necrotic).

In 2 out of the 21 WT and one out of the 2 MRTK diagnosed on CNB, the diagnosis was revised as CSSK on the final histology of nephrectomy specimen.(Table 2) The concordance between CNB and histology of resected specimen was 91%. The sensitivity, specificity, positive predictive value and negative predictive value of CNB in the

diagnosis of pediatric renal tumors were 89%, 50%, 89% and 100% respectively.

About three-fourths of the 19 excised surgical specimens had <33% viable tumor cells reported as the blastemal component, thus proving that the blastemal component is chemo-sensitive. Only 2 specimens had shown blastemal component of >66% thus designating it as a high-risk tumor with a poor prognosis.

DISCUSSION

In the field of pediatric oncology, obtaining an accurate diagnosis in the shortest period of time with minimal morbidity is of utmost importance.[17] Of all childhood cancers, malignant renal tumors comprise 7% of all cancers. Among them, WT is the most common renal tumor which is commonly presented in the age group from 1-4 years.[1,2,6] The differentiation between WT and NWRT can be challenging as they share many radiological features. Therefore, CNB is necessary for accurate diagnosis. UKCCSG WT Study 3 adopted pre-NACT CNB for histological diagnosis to address these issues [13].

The diagnosis of WT and other renal tumors is typically made by imaging studies, including USG (including Doppler), computed tomography and magnetic resonance imaging. These studies can provide detailed information about the size, location and characteristics of the tumor, which are invaluable in therapeutic decision making. In some cases, a CNB may be performed to confirm the diagnosis, but this is not always necessary, especially when the imaging studies are highly suggestive of a specific type of tumor.[5] According to Umbrella protocol, CNB is now considered only in cases with unusual clinical presentation like age >6 years, urinary infection or septicemia, psoas infiltration, pulmonary metastasis in <2 years, and extra-hepatic or extra-pulmonary metastasis, unusual radiological findings like numerous calcifications, voluminous lymphadenopathy, non visualization of renal parenchyma, extra-renal projections and biological findings like hypercalcemia or LDH level >4 x ULN (upper limit of normal).[18] It is our institutional policy to do CNB in every patient suspected to have a malignant renal tumor.

Skoldenberg's study showed that the sensitivity of CNB was 76%.[14] Taskinen *et al.* reported a sensitivity of 100% for CNB in diagnosing WT; however, only two out of five NWRT cases in their series were diagnosed with CNB.[4] Jackson *et al.* showed that the sensitivity and specificity of CNB were 86% and 99.6%, respectively. In 25 out of 518 cases, CNB changed the management, thereby demonstrating the value of this procedure.[15] Mitchell *et al.*[16] reported that approximately 12% of cases were diagnosed as NWRT due to CNB and were treated accordingly, even though they had features suggestive of WT in radiological studies.

CONCLUSION

We conclude that in our experience CNB is an effective tool in diagnosing WT and differentiating it from NWRT. Inappropriate NACT has been avoided in 2 patients who were diagnosed with NWRT on CNB. In 3 bilateral WT, NACT prescribed by SIOP Umbrella protocol didn't reduce any tumour volume, which indicates that we should consider administering 3-drug NACT as suggested in COG.

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Clinical Study

Morphology of Umbilical Hernia in Senegalese Children and Its Impact on the Clinical Presentation

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Keywords
Gandiaye classification
Umbilical hernia

Abbreviations
HIC - High-income countries
RUS - Redundant umbilical skin
UC - Umbilical cord
UH - Umbilical hernia

Abstract

Introduction: *Umbilical hernia is common in African children. However, studies on its morphology are scarce in sub-Saharan Africa. We aimed to characterize its morphology in Senegalese children and evaluate its impact on the clinical presentation.*

Methods: *We conducted a prospective analytic study for 6 months (March to September 2023) at the Albert Royer National Children's Hospital Centre in Dakar, Senegal. The association between parameters was studied with appropriate statistical tests.*

Results: *This study included 145 patients of umbilical hernia, of whom 121 (63.4%) were having normal overlying skin. A flat umbilicus was found in 16 patients (16%), and a conical umbilical hernia in 70 patients (48.3%). Medium-sized defects (0.5 - 1.5 cm) were found in 57 patients (39.3%), and redundant umbilical skin (RUS) was found in 111 patients (76.6%). The shape of the umbilical hernia and RUS was linked to the size of the fascial defect (p = 0.001 and 0.047, respectively). The 3 latter parameters were associated with the clinical presentation (p=0.026; 0.000 and <0.001, respectively). We found an association between the shape of an umbilical hernia and the extent of the RUS (p=0.000).*

Conclusion: *In Senegalese children, umbilical hernias frequently have normal overlying skin with a conical shape. However, umbilical hernia with a flat umbilicus is not exceptional. The fascial defect is usually medium, and redundant umbilical skin is present. Morphology of umbilical hernia is associated with its clinical presentation.*

INTRODUCTION

Umbilical hernia (UH) is estimated to occur in 25 to 58% of African children.[1] Its clinical presentation includes 3 types: simple UH (non-painful umbilical bulging as the only symptom), symptomatic UH (additional to the umbilical swelling, recurrent umbilical pain is reported), or complicated (which includes incarcerated and strangulated UH presenting with painful umbilical swelling, vomiting or obstipation with or without bowel ischemia respectively).[2] It is one of the commonest disease of pediatric surgical practice in Africa, where it is often symptomatic or complicated. It is significantly different from high-income countries (HIC), where complications are exceptional.[1,3,4] The higher rate of complications in African children resulted in recommendation against conservative management of UM as it is practiced in HIC. Some African authors suggest immediate repair of UH at diagnosis as it is done for inguinal hernia.[5,6]

The African literature on pediatric UH mainly covers its epidemiology, management, and outcomes.[7] The morphology of UH in African children is not adequately studied. Authors rarely give importance to it, such that only one report has studied it partially.[8] Many authors briefly allude to umbilical morphology while describing umbilical reconstruction (umbilicoplasty) after UH repair.[9]

Morphology of UH is important in the context of planning umbilicoplasty. Further, the morphology of UH was reported to impact the clinical presentation, that small and medium-sized defects being frequently encountered in complicated UH.[3] Therefore, we aimed to study the morphology of UH in Senegalese children by introducing a new classification that is easy to use and is based on the geometrical aspect of the umbilical swelling. Secondarily, we aimed to analyze the association between UH morphology and their clinical presentation.

PATIENTS AND METHODS

Study Setting and Design

We conducted a prospective analytical study for 6 months, from 17 March to 17 September 2023, at the Pediatric Surgical Department of Albert Royer National Children's Hospital Center. Established in 2010, it is one of the leading pediatric surgical departments in Senegal, managing patients from all regions of Senegal and neighboring countries such as Gambia, Guinea, and Mauritania.

Study Population

We included patients from 0 to 15 years of age diagnosed with UH during the study period whose parents or legal guardians freely consented to participate in the study. Helsinki Declaration of the World Medical Association was strictly observed. Patients presenting with recurrent UH were not included.

The study comprised of 145 patients, with a median age of 3 years (range 0.8 to 15 years). There were 94 males (64.8%) and 51 females (53.2%). About 20 patients had a medical comorbidity (13.8%) and 27 (18.6%) had a surgical comorbidity, of which 13 (9%) had an additional hernia (inguinal or epigastric). There were 84 patients (57.9%) with simple UH, 47 (32.4%) with symptomatic UH and 14 (9.7%) with incarcerated UH.

Data Collection

Senior Residents of our department physically examined patients using a measuring tape and a marker. Patients were examined in standing and in dorsal decubitus positions. The aspect of the skin overlying the hernia was noted on inspection in either position. For measuring the defect size, the patient was made to lie down on dorsum, the hernia was reduced and the examiner's fingers located the defect's margins which were marked using a skin marking pen. The marked distance was then measured using a scale. (Fig. 1) For measuring the redundant umbilical skin (RUS), the patient still lying on dorsum, the hernia

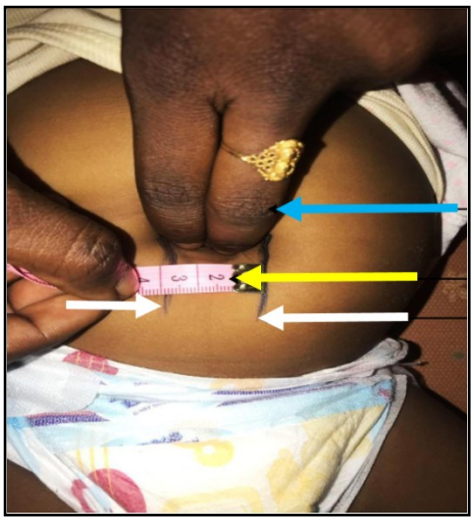

Fig. 1: Technique of measuring the fascial defect
The defect is delineated by fingers (blue arrow) and marked with ink (white arrows). Using a measuring tape (yellow arrow) the marked defect is measured

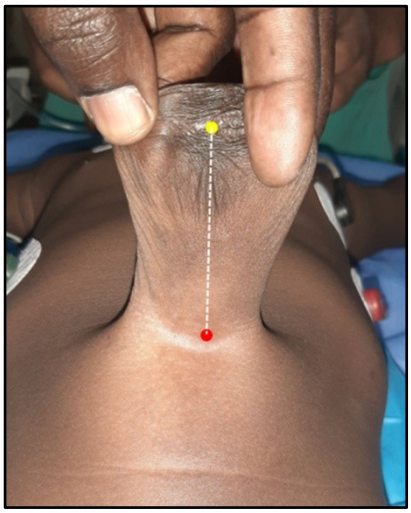

Fig 2. Measurement of Redundant Umbilical Skin
After reducing the umbilical hernia, the apex of the hernia is identified (yellow dot) as well as the lowest point of the lower umbilical crease (red dot). The distance between these two dots (interrupted white line) represents the length of redundant umbilical skin

was reduced, and two points were marked on the apex and the lowest point of the lower umbilical crease.(Fig. 2) With a measuring tape, the distance between these two points was measured as RUS. For recording the shape of the hernia, the patient was standing up without reducing the hernia. In children who were too young to stand on their own, the parents held them in an erect position. We devised a new *Gandiaye classification*, which categorizes the shape of UH depending on the geometrical aspect (conical, ellipsoidal, ovoidal, and spherical). These data were registered on a predesigned questionnaire and encoded on an Excel (Microsoft Office TM 2020) spreadsheet.

Study Variables

The parameters studied include: (a) appearance of the overlying skin (normal, hyperchromic or hypochromic), (b) defect size, categorized based on *Lassaletta's classification* (small, medium, and large defect)[10], (c) shape of the hernia, using the *Gandiaye classification of UH* (conical, ellipsoidal, ovoidal, and spherical), and (d) RUS (0.5 - 1.5 cm, and 1.6 - 5 cm).

Statistical Analysis

Analysis was performed using Statistical Package for Social Sciences software (IBM SPSS version 26). Categorical variables were presented as frequencies and continuous variables as mean (with standard deviation) or median (with first and third quartiles) depending on their Gaussian distribution. The relationship between UH defect size and overlying skin, UH shape, and RUS was accordingly evaluated using Pearson's chi-squared test and Monte Carlo exact test. The impact of UH morphology on clinical presentation was studied by comparing the morphological parameters in the 3 clinical presentation groups, using the same statistical tests accordingly. Statistical significance was set at P - value<0.05.

RESULTS

This study enrolled 145 patients. Of these, 121 patients (83.4%) had normal overlying skin color, while 20 patients (13.8%) had hyperchromic skin, and 4 (2.8%) had hypochromic skin. (Fig 3) The median fascial defect size was 1 cm (Range 0.7 – 2 cm).

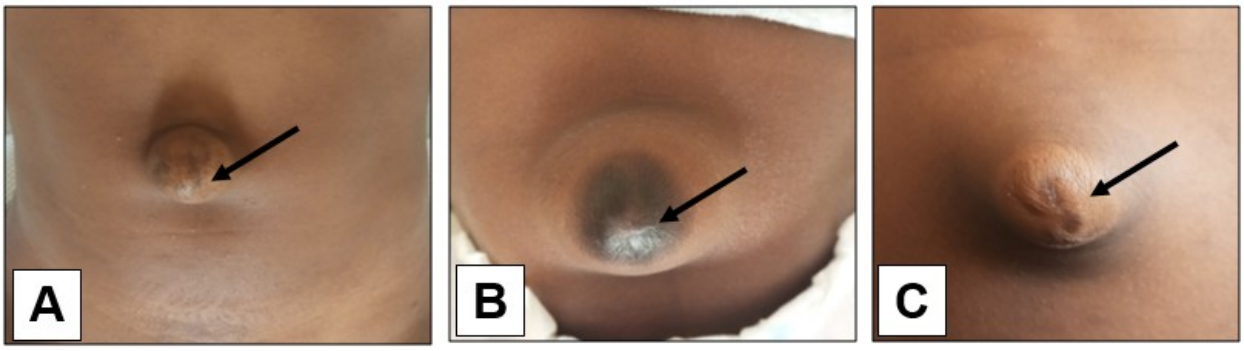

Fig 3. Overlying skin of umbilical hernia.
Skin colour is (A) normochromic, (B) hyperchromic and (C) hypochromic

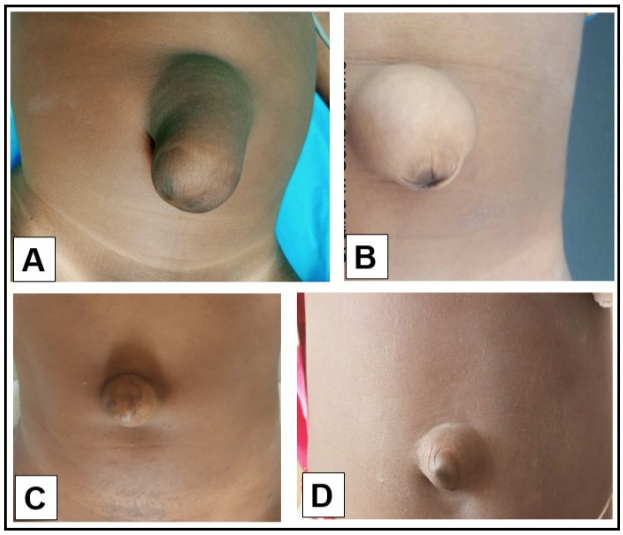

Fig 4. Gandiaye classification of umbilical hernia
(A) Conical shape, (B) Ovoidal shape, (C) Spherical shape, (D) Ellipsoidal shape

Table 1. Shape of the umbilical hernia (The *Gandiaye classification*)

Shape of Hernia	Number	Percentage
Flat umbilicus	16	11
Elevated umbilicus	82	89
Conical	70	48.3
Ovoidal	24	16.4
Ellipsoidal	24	16.4
Spherical	11	7.6

According to the Lassaletta's classification, small-sized defect (< 0.5 cm) was found in 46 patients (31.7%), medium-sized defect (0.5 – 1.5 cm) in 57 patients (39.3%), and large-sized defect (> 1.5 cm) in 42 patients (29%).

Sixteen patients (11%) had flat umbilicus. In contrast, 129 others (89%) presented with an elevated umbilicus with different geometrical aspects, as reported in Table 1. (Fig 4)

The median size of RUS was 1 cm (Range 0.5 –5 cm). About 34 patients (23.4%) had no RUS; it was found in 111 others (76.6%). Among the latter, it ranged from 0.5 to 1.5 cm in 78 patients (70.3%), from 1.5 to 5 cm in 33 patients (29.7%), and none had more than 5 cm. Analysis of the relation between the defect size and aspect of the overlying skin, UH shape, and RUS is represented in Table 2. Flat umbilicus and spherical UH were more frequent associated with small defect size, ovoidal UH with increased defect size and ellipsoidal UH with medium-sized defects. RUS proportionally increased with the defect size.

In Table 3, the impact of UH morphology on clinical presentation is depicted. We noticed that UH with a flat umbilicus was mainly symptomatic while the other shapes tended to present mainly as simple UH. Small-size defects tend to be more symptomatic, while medium and large-size defects usually presented as simple UH. Additionally, the frequency of incarcerated and symptomatic UH decreased with increasing defect size. Umbilical hernias without RUS were mainly symptomatic, while those with RUS were often simple UH. When UH with a flat umbilicus naturally does not have RUS, spherical UH all had RUS ranging from 0.5 to 1.5 cm. (Table 4)

Table 2. Clinical features of umbilical hernia

Defect size	Small *n* (%)	Medium *n* (%)	Large *n* (%)	p-value
Skin Colour				0.65 ‡
Normal	41(33.9)	46 (38.0)	34 (28.1)	
Hyper chromic	4 (20)	10 (50)	6 (30)	
Hypo chromic	1 (25)	1 (25)	2 (25)	
UH shape				<0.001 ‡
Flat	11 (68.8)	5 (31.2)	0	
Conical	21 (30)	27 (38.6)	22 (31.4)	
Ovoidal	3 (12.5)	8 (33.3)	13 (54.2)	
Ellipsoidal	6 (25)	11 (45.8)	7 (29.2)	
Spherical	5 (45.5)	6 (54.5)	0	
RUS				0.047 †
Absent	17 (50)	11 (32.4)	6 (16.4)	
0.5 – 1.5	21 (26.9)	35 (44.9)	22 (28.2)	
1.5 – 5	8 (24.2)	11 (33.3)	14 (42.5)	

RUS: Redundant umbilical skin
‡Monte-Carlo Exact test, †Pearson's chi-square test

DISCUSSION

The umbilicus is the only natural scar in the human body that plays an esthetic role. Hence a sizable number of patients with UH present because of esthetic concerns.[11] However, in Africa, majority of patients present with painful episodes, which led UH to be extensively studied in this continent.[3,4,6,12] However, no study was entirely dedicated to its morphology, as most of them partially discussed that aspect when studying umbilicoplasty after UH repair.[9,11,13] In this study, 8 in 10 patients presented with normal color of the skin over the hernia, while a tenth presented with hyperchromic skin. However, the aspect of the overlying skin did not show an association with the defect size or the clinical presentation. We hypothesize that this could be linked to the width of the umbilical cord (UC). Large size UC would need more dressings, resulting in a hyperchromic scar, while normal-sized UC would allow rapid healing with normal overlying skin.[14,15] Hypochromic skin over the hernia can be explained by a deep wound after umbilical cord separation or early exposure to sun rays.[15]

Based on the report of Lassaletta et al. in 1975[10], the size of fascial defect in UH is categorized into three types. They observed that small-size defects may spontaneously close more rapidly than larger ones, in which spontaneous closure could happen around four years or more.[10] Therefore, in HIC, the defect size is used to predict the time to spontaneous closure.[16] In our study, the smaller the defect, the higher the frequency of symptomatic or incarcerated UH. In African children, high frequency of complications prompted extensive studies on their risk factors, among which our findings on the size of the fascial defect confirmed the association of smaller size defects with more complications.[3,8,17–18] For this reason, some authors suggested routine early repair of small and medium-sized defect UH.[5, 6] In contrast to the practice in HIC, smaller defect size in African children prompts early repair of UH.

An UH can be of various geometric shapes. In this study, conical UH was the most common, followed by ovoidal, spherical, and ellipsoidal shapes. A tenth of our patients had a flat, normal-looking umbilicus. This emphasizes that UH can occur in children with normal-looking umbilicus, which is found exclusively in small-size defects. Therefore, in African children, systematic umbilical palpation should be performed even in well-looking child complaining of abdominal pain, which may be due to herniation of the omentum.[2] Additionally, the shape of UH was associated with the clinical presentation. It means, the clinician could predict the occurrence of complications or symptoms based on the defect size and shape. Umbilical hernias with flat umbilicus should be operated early as they are mainly symptomatic.

RUS adversely affects the esthetic appeal of the umbilicus.[9,10] The degree of RUS is linked to the size of the fascial defect; the wider it is, the more

Table 3. Impact of UH morphology on clinical presentation

Clinical Presentation	Incarcerated *n* (%)	Simple *n* (%)	Symptomatic *n* (%)	*P*-value
Skin Colour				0.484 ‡
Normal	14 (11.6)	68 (56.2)	39 (32.2)	
Hyperchromic	0	14 (70)	6 (30)	
Hypochromic	0	2 (50)	2 (50)	
UH shape				0.026 ‡
Flat	0	5 (31.2)	11 (68.8)	
Conical	11 (15.7)	42 (60)	17 (24.3)	
Ovoidal	0	16 (66.7)	8 (33.3)	
Ellipsoidal	3 (12.5)	14 (58.3)	7 (29.2)	
Spherical	0	7 (63.6)	4 (36.4)	
Defect size				0.000 ‡
Small	8 (17.4)	15 (32.6)	23 (50)	
Medium	5 (8.8)	32 (56.1)	20 (35.1)	
Large	1 (2.4)	37 (88.1)	4 (9.5)	
RUS				<0.001 ‡
Absent	1 (2.9)	12 (35.3)	21 (61.8)	
0.5 – 1.5	7 (9)	53 (67.9)	18 (23.1)	
1.6 – 5	6 (18.2)	19 (57.6)	8 (24.2)	

RUS: Redundant umbilical skin, ‡ By Monte-Carlo Exact test

Table 4. Relationship between the shape of umbilical hernia and RUS

RUS	Absent *n* (%)	0.5-1.5 cm *n* (%)	1.6-5 cm *n* (%)
UH shape			
Flat	16 (100)	0	0
Conical	4 (7.1)	41 (58.6)	24 (34.3)
Ovoidal	1 (4.2)	15 (62.5)	8 (33.3)
Ellipsoidal	12 (50)	11 (45.8)	1 (4.2)
Spherical	0	11 (100)	0

‡- P-value calculated by Monte-Carlo Exact test is <0.0001
RUS: Redundant umbilical skin

abdominal viscera would pass through and cause stretching of the umbilical skin. Since the skin is elastic, RUS gets extended as long as the defect remains wide. This is reinforced by the observation that RUS is more common in simple UH, as symptomatic and incarcerated UH mainly occur in small and medium-size defects.[3] Concerning the cosmetic aspect, we believe that assessing RUS is as important as assessing the size of the fascial defect. Knowing the degree of RUS is essential in planning umbilicoplasty as to which technique to be used. Usually, an umbilicoplasty is needed if RUS exceeds 1.5 cm.[9] Based on that, spherical and ellipsoidal UH do not need umbilicoplasty, while a third of conical and ovoidal UH will need it. However, a definitive indication of umbilicoplasty should be made after the closure of fascial defect and tacking the dermis to the fascia during the UH repair. Some UH with considerable RUS may not finally require an umbilicoplasty after these steps.(Fig 5) Further, the long-term benefits of umbilicoplasty is still debatable as umbilicoplasty in UH with RUS uses flaps, which leave an additional scar and some additional complications such as surgical site infection, keloid, and stitch granuloma.[9] It is known that umbilical shape in children keeps varying up to adolescence and hence patient satisfaction with umbilical cosmesis after umbilicoplasty may also vary with age.[20]

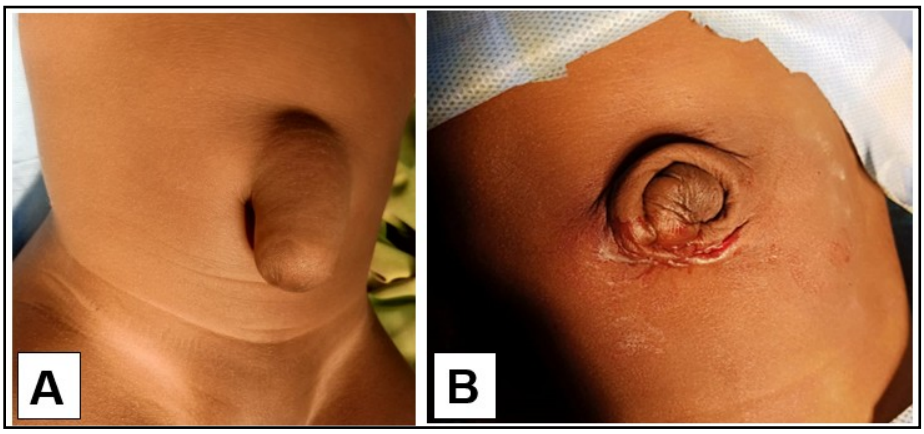

Fig 5. Cosmesis of umbilical hernia repair

(A) Presence of redundant umbilical skin may cause to consider the inevitability of an umbilicoplasty. However, (B) after fascial closure and dermis tacking to the fascia, redundant skin is less apparent, thereby obviating the need for umbilicoplasty.

Future studies must evaluate the subjective satisfaction of umbilical shape in children who underwent UH repair.

CONCLUSION

Morphology of UH in Senegalese children is dominated by medium-sized, conical shaped hernia with normal-looking overlying skin colour and RUS. In some children UH presented with a normal-looking umbilicus. This emphasizes the necessity of umbilical palpation in children complaining of recurrent abdominal pain. The defect size, umbilical shape, and extent of RUS were associated with the clinical presentation of UH.

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Clinical Study

Posterior Urethral Valve: Is Complete Endoscopic Valve Incision Sufficient?

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Keywords
Endoscopic valve incision
Posterior urethral valve
Urethral ratio

Abbreviations
APD - Antero-posterior diameter of the renal pelvis
BNI - Bladder neck incision
CRF - Chronic renal failure
ESRD - End-stage renal disease
EVI - Endoscopic valve incision
MCUG - Micturating cysto-urethrogram
PTK - Parenchymal thickness of kidney
PUV - Posterior urethral valve
PVRV - Post-void residual volume
UDS - Urodynamic study
UR - Urethral ratio
UTI - Urinary tract infection
VUR - Vesico-ureteric reflux

Abstract

Background: *After adequate endoscopic incision of posterior urethral valve (PUV), normalization of posterior urethra on micturating cysto-urethrogram (MCUG) is expected. This study examines if normalization of urethra is correlating with check urethro-cystoscopy, improvement of renal function tests, imaging of upper renal tracts and urodynamics studies in all patients.*

Methods and Material: *Fifteen consecutive patients with PUV from June 2011 to May 2012 were included in this prospective study. Renal function tests, ultrasonography and MCUG were done in all patients at diagnosis. Urethral ratio (UR) was calculated by dividing posterior urethral diameter with anterior urethral diameter in oblique MCUG films. However, it was seen that each MCUG yields a range of UR and hence we labelled the least UR as the 'best UR', and the maximum one as 'worst UR'. Pre-operative urodynamic studies were also done in these patients. All the investigations were repeated 12 weeks after adequate endoscopic valve incision (EVI). All the patients were assessed for the need of any further management. Statistical significance was calculated using Mann-Whitney U-test.*

Results: *The median age at presentation was 21 months (range 8 d - 7 yr). Pre and post EVI values of renal function tests did not differ significantly. Reduction of hydronephrosis and VUR was not significant; but reduction of bladder wall thickness and hydroureter was significant. The mean pre-EVI 'best UR' was 3.58 (range 1.6 - 8.3; SD 1.86) and the mean post-EVI 'best UR' was 1.7 (range 1 - 3.3; SD 0.61). This difference was significant ($p<0.001$). Post-EVI urodynamic abnormalities were seen in 71.43% of patients in whom urodynamic study (UDS) was done. Two thirds of patients needed some form of bladder management even after adequate EVI.*

Conclusions: *Following adequate EVI, there is a significant reduction in the 'best UR'. Adequate EVI however does not correlate with improvement in renal imaging, renal function tests and urodynamic parameters.*

INTRODUCTION

Posterior urethral valves (PUV) are the most common cause of lower urinary tract obstruction in male infants, with an incidence ranging from 1/3000 to 1/8000 male births.[1] It is now agreed that primary endoscopic valve incision (EVI) is the treatment of choice for PUV. One-third of boys with PUV progress to end stage renal disease (ESRD).[2] Some surgeons believe that progression of bladder dysfunction and the ultimate ESRD are the results of inadequate endoscopic treatment of PUV. They contend that if valve incision were adequate, further progression of renal and bladder dysfunction would not take place.[3,4] Another school of thought suggests that the basic pathology is an embryological field defect in the urinary tract, which persists despite adequate EVI. According to this school, bladder is the most affected organ in this field defect and hence proper bladder management would preserve the upper tracts in most of these patients. Some workers believe that patients presenting with gross upper tract dilatation, highly deranged renal function tests and urosepsis, would ultimately land up in ERSD. This may be because of teratogenic insult not only at the site of the origin of Wolffian ducts, but also at the site of nephrogenesis.[5]

In the past, adequacy of treatment was judged by surgeons by mere inquiries and observation of the urinary stream. Check cystoscopy has always been considered as the gold standard for documentation of complete ablation of valves.[6] Recently, some authors have stated that adequate EVI is indicated by post procedural normalization of the UR.[3,7] However, there are differing opinions as regard to the correlation of these changes with the changes in the postoperative general condition, voiding status, improvement in renal function and drainage patterns of the upper urinary tract.[3,6]

In this study, we aimed to evaluate whether adequate EVI alone, as suggested by normalization of urethral ratio on MCUG and confirmed by check urethro-cystoscopy, is also associated with improvement in bladder function, upper urinary tracts and renal function tests in PUV patients.

MATERIALS AND METHODS

Fifteen consecutive patients with classical PUV managed in the Department of Pediatric Surgery over a period of one year (from June 2011 to May 2012) were included in this prospective study. Renal function tests and ultrasonography was done in all of them at diagnosis. Those with urinary tract infections (UTI) were treated with suitable antibiotics and catheterisation.

Urethral ratio (UR) was calculated in all the patients by dividing posterior urethral diameter with anterior urethral diameter in oblique MCUG films. The diameter of posterior urethra was measured transversely at a point half way between the bladder neck and the distal end of membranous urethra. The transverse diameter of anterior urethra was measured at the point of maximum distension at bulbar urethra. We realized early in the study that the UR varies with changing dimensions of the anterior urethra in correlation with the changes in voiding pressure during different phases of micturition. So, a range UR was available for each MCUG. We labelled the one with least value as 'best UR' and the maximum one as 'worst UR'. (Fig. 1) The difference between 2 values were analysed by using paired sample t-test. For 7 MCUGs, only a single micturating film was available and hence instead of two values of UR, only one value was available in them. Pre-operative urodynamic studies (UDS) were also performed in 5 patients.

PUV were endoscopically incised using a hook electrode through an appropriate size urethro-cystoscope at 5, 7 and 12 o'clock positions. UR was recalculated on MCUG performed 12 weeks post procedure in the same fashion. Two criteria were to be fulfilled to label an adequate EVI: (1) Post-EVI, UR of <3 was considered as radiological normalisation of posterior of urethra; and (2) evidence of adequate valve incision at check urethro-cystoscopy. If either of these two criteria

were not met, EVI was done once again. If adequate treatment of PUV was ascertained, renal function tests, ultrasonography and UDS were done. UDS was not done in infants (n=7), in those with high-grade VUR (n=4) and 2 other patients in whom pre-EVI catheterization was not successful.

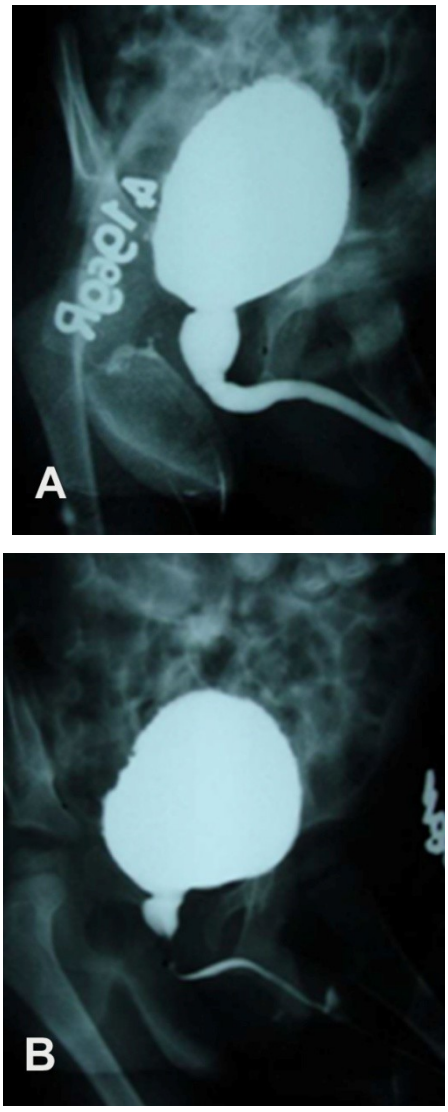

Fig. 1: Concept of 'best' and 'worst' urethral ratio
MCUG showing the 'best' (A) and 'worst' (B) urethral ratios in the same patient after endoscopic valve incision

Patients with persistently abnormal bladder functions were considered to be the candidates for treatment with anti-cholinergics and/or bladder neck incision (BNI).

Mann-Whitney U-test was done to evaluate the statistical significance of the difference noted between the pre- and post-ablation values of blood urea, serum creatinine, antero-posterior diameter of renal pelvis (APD), parenchymal thickness of kidney (PTK), diameter of ureters and bladder wall thickness. Same test was used to evaluate the statistical significance of difference between pre- and post-ablation values of 'best UR'. P - value < 0.05 was considered significant.

RESULTS

The median age at presentation was 21 months (range of 8 days to 7 years). There were 4 neonates, 3 infants, 5 were between 1-5 years, and 3 were pre-schoolers. Only 2 patients were antenatally diagnosed, while 2 presented with febrile UTI. All others had obstructive urinary symptoms.

The mean pre-EVI blood urea was 31.8mg/dl (range 12-66 mg/dl; SD 15.58) and the mean post-EVI blood urea was 30.67 mg/dl (range 16-72 mg/dl; SD 16.27). The mean pre-EVI level of serum creatinine in these patients was 0.8 mg/dl (range 0.3- 2.5 mg/dl; SD 0.63) and the mean post-EVI serum creatinine was 0.53 mg/dl (range 0.1- 1.3 mg/dl; SD 0.29). Pre- and post-EVI values of blood urea and serum creatinine were not found to be statistically different ($P = 0.42$ and 0.83 respectively). Only 1 patient had post-op creatinine of more than 1 mg/dl. Pre- and post-EVI PTK was available for 26 renal units, APD in 24, ureteric diameter in 21 and bladder wall thickness in 11 patients. The mean improvements in pre- and post-EVI values of these sonographic parameters are shown in Table 1.

By Mann-Whitney U-test pre- and post- EVI values of PTK and APD did not differ significantly (P=0.4 and 0.35 respectively). However, sonographic parameters of the 21 ureters and 11 bladders showed significant difference between the pre- and post-EVI values (P=0.003 and 0.024 respectively).

Post-void residual volume (PVRV) was significant in 6/15 (40%) patients before treatment and it was significant in only 2/15 (13.33%) of patients after adequate valve ablation. By applying the Fisher's exact test, we found this difference was not statistically significant (P=0.21).

Table 1. Ultrasound findings in PUV patients

Parameter	n	Mean percentage improvement	SD %	*P* value†
PTK (mm)	26*	11.02	41.97	0.41
APD (mm)	24*	11.15	49.53	0.35
Ureteric diameter	21*	13.37	23.90	0.003
Bladder wall thickness	11	15.66	21.81	0.024

** Numbers indicate renal / ureteric unit*
† Calculated by Mann-Whitney U- test.
PTK- Parenchymal thickness of kidney; APD-Antero-posterior diameter of renal pelvis

The mean pre-EVI 'best UR' was 3.58 $\pm$ 1.86 (range 1.6-8.3). The mean post-EVI 'best UR' was 1.7 $\pm$ 0.61 (range 1-3.3). After analysing the two set of values, the difference was significant (P<0.001). (Table 2)

VUR was seen in 4/30 (13.3%) ureteric units before treatment, with two patients having unilateral grade-V VUR and one with bilateral grade-V VUR. Five out of 30 (16.67%) units had VUR after adequate EVI. The patient who had bilateral grade-V VUR at presentation had complete resolution of VUR post-EVI, while those with unilateral grade-V VUR at presentation showed no change; instead one of these developed grade-V VUR on the contralateral side too. Two patients with no VUR at presentation developed unilateral VUR later, grade III and V (one each). Most (80%) of patients needed single procedure for adequate EVI by our criteria, while a repeat EVI was needed in 2 patients and 4 times in 1 child.

Table 2: The pre- and post-EVI 'best urethral ratios'

Pt. No.	Pre-EVI	Post-EVI	Decrease† (%)
1	3.4	2.2	35.29
2	3.5	1.9	45.71
3	1.6	1	37.5
4	2.5	1.6	36
5	2.1	1.5	28.57
6	3	1	66.67
7	6	2	66.67
8	8.3	1	87.95
9	5.6	3.3	41.07
10	2.5	2	20
11	2.4	1.3	45.83
12	2.6	1.6	38.46
13	2.5	1.4	44
14	2.5	2.3	8
15	5.2	1.5	71.15

(Mann-Whitney U-test, p-value < 0.001)
EVI- Endoscopic valve incision
†Calculated by the formula: difference between pre and post EVI ratios/Pre-EVI ratio x 100

Urodynamic studies were done in 5 pre-EVI and in 7 post-EVI patients. Comparison of pre- and post-EVI UDS findings was available in only 5 patients. (Table 3) Post-EVI UDS was done in 2 more patients in whom pre-EVI UDS was not possible. One patient out of these 2 had normal study and another had overactive bladder. Five patients developed episodes of febrile UTI during the course of study and they were all managed conservatively.

We finally assessed all outcomes in our study individually for every patient to decide whether any further treatment was needed in them or not. The further treatments required by our patients were oxybutynin (n=5) and bladder-neck incision (BNI) (n=3). Indications for oxybutynin were overactive bladder, low capacity bladder and progression of VUR with trabeculated bladder on MCUG. Indications for BNI were bladder neck hypertrophy, back-pressure changes in the bladder on MCUG or check urethro-cystoscopy and significant PVRV after adequate EVI. BNI was

combined with oxybutynin medication in 2 patients. Surprisingly, the only patient with chronic renal failure (CRF) in our study is doing well on CRF medications alone. Four other patients are also doing well and required no further treatment until this date. Out of these 4 patients, one has dilated cardiomyopathy and is also undergoing treatment for that.

Table 3: Outcome of urodynamic studies

Sr. No.	Bladder status	
	Pre-EVI	**Post-EVI**
1	Low capacity high pressure	Same as pre EVI
2	Low capacity	Same as pre EVI
3	Low capacity high pressure	High pressure overactive
4	High pressure	Normal study
5	High pressure	Overactive bladder

DISCUSSION

Although it has been well documented that bladder management following EVI is the most important aspect of PUV management, there are reports claiming that adequate EVI is sufficient in most of them and inadequate EVI is the chief cause of progressive damage of the upper tract.[1-4, 8-15] Also, opinions differ regarding the optimum investigation for the confirmation of adequate EVI. While some authors consider the repeat MCUG to be sufficient, others consider check urethro-cystoscopy as the gold standard. This study was an attempt to find whether adequate EVI alone is sufficient in PUV patients.

Prior to this study, UR has been described in 3 papers in the literature. One study[1] observed post-EVI UR of 3 or less as a marker of adequate EVI, while a cut-off of 2.5-3 was suggested by another study.[7] Yet another study emphasized the importance of the UR calculated 3 months after EVI and observed that those within 2 standard deviations of their post-EVI UR fared better than others and required no further treat-ment. However, they did not mention any cut-off value as marker of complete EVI.[3] Smeulders retrospectively reviewed 31 patients.[6] Repeat MCUG was suggestive of residual valves in 10, but no residual leaflets were identified on check urethro-cystoscopy in 4. In 20 patients, the valves that appeared completely ablated on MCUG had persistent valve leaflets on cystoscopy and they required further ablation in 10. They concluded that positive predictive value of repeat MCUG was 56% and negative predictive value was 50%.

In our study, post-EVI value of mean+2SD of best UR was 2.93 and that of mean+1SD was 2.32. So, our results matched with the observations of Gupta.[7] Although our observation of difference of pre- and post-EVI 'best UR' was statistically significant, 8 (53.33%) of our patients had UR less than 3 at presentation. So we could infer from our findings that UR, though was a good marker of adequate EVI in our study, it was not a good diagnostic tool. Also, as 14/15 patients in our study had maximum post adequate EVI, UR of less than mean+1SD (2.32), we consider 2.3 as a good cut-off value. Our observation of 'best' vs. 'worst' UR emphasizes the importance of calculating UR at the time of peak flow as the distension of anterior urethra is maximum at that point of time. So, it is very important to use fluoroscopy which can freeze a frame from the recorded act of voiding. Video-urodynamic study is perhaps the most ideal method.

Smith *et al.* observed that the incidence of CRF increases with age.[8] In their study of long-term follow-up, 34% had CRF at 10 years of age, which increased to 51% at the age of 20 years. However, in most of their patients bladder management strategy constituted only of double voiding. In our study, 4/15 (26.67%) patients had serum creatinine of more than 0.8mg/dl at presentation, which decreased to 1/15 (6.67%) after adequate EVI.

We studied change in hydronephrosis and hydroureter separately. We observed statistically significant reduction in hydroureter following

adequate EVI, but not in hydronephrosis. We can say from our observation that the resolution of hydroureter precedes that of hydronephrosis, after EVI. Dunohoe *et al.* found persistent hydronephrosis in 20/71 (28.2%) patients in their study.[17] However, their follow-up period in these cohort was variable. They further performed video-urodynamics and started some form of bladder management in all these patients. On subsequent follow-up, there was further improvement and only 7 patients had residual hydronephrosis. Extrapolating these findings to our study, we can say that further follow-up after early establishment of appropriate bladder management in our series would yield us similar outcomes.

Most of the investigators have identified bladder dysfunction as the most important cause for worsening of CRF with age.[9-14] Parkhouse *et al.* identified four factors responsible for poor outcome: presentation before 1 year of age, proteinuria, bilateral VUR and day-time urinary incontinence. They were probably the first to speculate that bladder dysfunction even after resolution of obstruction is a major determinant of bad outcome.[9] Role of bladder dysfunction in ESRD has been stressed in the literature.[10-14] Different studies have observed bladder dysfunction on UDS in 50-90% of PUV after adequate EVI.[15,18,19] In this study, we found that overall bladder dysfunction was present in all patients at presentation. This improved by only 20% after adequate EVI. We did post EVI UDS in 7 patients in our study and observed bladder dysfunction in 5 patients. So, based on our UDS findings, 71.43% of patients needed attention of bladder dysfunction even after adequate EVI. Common abnormalities observed in our study were low bladder capacity, high pressure bladder and detrusor overactivity. This was in contrast to the observations made by Menon *et al,* according to whom most of the patients do not need any further treatment after adequate EVI.

In our study, PVRV was significant after adequate EVI in 2 (20%) patients aged more than 1 year and in none aged less than 1 month. Pre- and post-EVI difference in PVRV was not statistically significant (P=0.21). Incidence of significant PVRV after EVI has been variously reported in the literature, ranging from 12.5 to 81.43%.[20-22] Abraham *et al*. found significant improvement in PVRV after administration of terazosin following EVI.[23] One of our previous study showed encouraging improvement in bladder function after BNI; however, it was statistically insignificant. This could be a fallacy due to a small sample size.[24] We continue to perform BNI in patients with significant PVRV after EVI.

Ten out of 15 (66.67%) of our patients needed some form of bladder management strategies even after documentation of complete EVI. We applied multivariate logistic regression for all the biochemical and radiological parameters between those patients who needed some treatment after adequate EVI and those who did not. We observed that no parameter was statistically significant and thus no single parameter was indicative of the need for further treatment.

We believe in aggressive management of any bladder abnormality in our follow-up patients of PUV. Once we have documented complete EVI on check urethro-cystoscopy, we perform regular MCUG and UDS in these patients (besides USG and renal function tests) to promptly address any issue. Based on this principle, we observed that two thirds of our patients in this study needed some form of bladder management after 3 months of adequate EVI. We are of the opinion that the remaining one third of the patients in our series may also need bladder management in the future. We will keep a close follow-up of all the patients in our study and take prompt actions as and when required.

There were a few limitations in this study. A larger sample size and longer follow-up are needed to

make meaningful conclusion. This could not be achieved as the study duration was only one year. It is difficult to predict as to how much time is required for voiding dysfunction and other parameters to improve after successful EVI. For these reasons, we tried to assess the improvement after 12 weeks of adequate EVI. UDS was not done in infants because of technical difficulties. UDS was also not done in patients with high grade VUR as it was fallacious without video-urodynamics, which was not available in our center.

CONCLUSIONS

UR is not a static figure on a particular MCUG. A single MCUG would produce a range of UR and hence we propose to coin the terms 'best UR' and 'worst UR'. Cut-off values for 'best UR' of 2.5-3 should be considered as a marker of adequate EVI. Resolution of hydroureter precedes that of hydronephrosis in these patients. Two thirds of the patients need some form of bladder management even after adequate EVI and hence UR, though a good marker of adequate EVI, is not a indicator of completion of therapy in these patients. Therefore, we conclude that adequate EVI measured by normalization of UR may not improve renal function tests, imaging of upper renal tracts and urodynamics studies in all patients of PUV. Most of these patients may need some form of bladder management after EVI.

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Invited Expert Commentary

Unraveling Noma in the Modern Age

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Abstract

(David Allen Shaye is a leading expert in the reconstruction of mutilating defects caused by noma. In this invited commentary, he is sharing the wisdom and knowledge that he gained from his extensive work in Noma Children's hospital, Nigeria).

Management of noma patients is a surgical challenge not only because it occurs in low-income countries, but also because it is an infinitely complex, multi-faceted surgical problem. It is a peculiar amalgamation of oncologic, congenital, and traumatic facial defects. As there is no noma surgeon experienced in tackling all of these issues, noma lends itself to team-based care. The roots of noma are neither surgical, nor even medical, but a societal problem of the fair distribution of basic resources. Today, noma remains a stark reminder of our global failures, but tomorrow it could be the mark of a global triumph.

As surgeons we become familiar with death and disability; so it is the rare malady that shakes our stoic front. Noma is just this disease. A haunting gangrene of skin, muscle and bone, noma afflicts our most vulnerable population, namely the malnourished children of low-income countries. Survivors of noma are few, and they present to us a surgical challenge of the highest order.

In this (April 2024) issue of *Pediatric Surgery in Tropics*, Srinidhi and Raveenthiran have presented an outstanding assessment of this little-known disease, noma (cancrum oris, orofacial gangrene or necrotizing stomatitis).[1] This article comprehensively reviews the history, microbiological profile, layperson and medical nomenclature, and surgical principles of the disease, in an open access format that maximizes its global reach. The existing evidence on noma is woven together into a thorough narrative, excellent for all levels of health-care professionals. For those interested in the psychosocial effects of noma that is easily lost in the medical literature, *'Restoring Dignity'* is a riveting documentary of noma patients set in northwest Nigeria.[2]

Management of noma patients is a surgical challenge not only because it occurs in low-income

countries, but also because it is an infinitely complex, multifaceted surgical problem. In my experience, noma is a peculiar amalgamation of oncologic, congenital, and traumatic facial defects. The substantial loss of tissue during acute noma is veiled when the patient presents years later. As noma survivors grow, their facial skeleton and soft tissues are distorted comparable to congenital facial clefts, and the dentate facial skeleton is altered like that of neglected trauma. As there is no noma surgeon experienced in tackling all of these issues, noma lends itself to team-based care. The time is ripe for an international surgical working group to describe and lay out the principles of acute noma treatment, noma surgery and anesthesia.

In 2023, the World Health Organization (WHO)[3] recognized noma as a *neglected tropical disease*. This brings hope for an accelerated, integrated approach, particularly within the hardest hit continent of Africa. A global strategy including epidemiology, community health, and specialty care is needed. The review of risk factors shows us the many confounding variables that contribute to the unsolved nature of the disease. The hope to discover a single risk factor as a chokepoint for eradication remains elusive. However, with the common theme of noma being severe malnourishment and poverty, perhaps these focal points do not require a deeper understanding. The roots of noma are neither surgical, nor even medical, but a societal problem of the fair distribution of basic resources. Today, noma remains a stark reminder of our global failures, but tomorrow it could be the mark of a global triumph.

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